

Ectopic Meningioma of Frontoethmoid Region

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Abstract

Meningiomas arise from arachnoid cells. They can occur at intracranial or extracranial sites. Extracranial meningiomas are very rare tumors. They may arise from nasal cavity, paranasal sinuses, middle ear, and subcutaneous tissues. The ectopic meningiomas of paranasal sinuses do not have any communication with intracranial meninges. They usually occurs in young males. Imaging studies shows no bone erosion or intracranial extension. Primary surgical excision is the treatment of choice. The present case is reported due to its rarity and presentation at an unusual age.

Keywords: Ectopic meningioma, meningioma of paranasal sinus, paranasal sinus tumor.

INTRODUCTION

The term “meningioma” was coined by Cushing to describe the tumor that arises from central nervous system meninges.¹ Meningiomas are neoplasms derived from arachnoidal (meningothelial) cells. Meningiomas accounts for approximately 18% of all primary intracranial neoplasms and 25% of intraspinal neoplasms.²⁻⁴ Meningiomas outside the CNS are uncommon. They can be primary/ectopic (occurring at ectopic site) or secondary (extending from an intracranial lesion). The common sites of ectopic meningioma are skull bones, paranasal sinuses, nose, orbit, scalp, middle ear, neck and skin.⁵ The ectopic meningioma of skull is a rare tumor accounting for about 1% of meningiomas. They are derived from the embryonic arachnoidal cell remnants of ectopic meningocytes derived from pluripotent mesenchyme cell captured at inappropriate site during embryonic development or implantation of these cell by same mechanical insult like trauma or dural tear.⁶ Intradiploic meningioma of the frontoethmoid orbital complex is extremely rare and only 10 cases have been reported till date.⁶⁻⁸

CASE REPORT

A 45 years old male presented with complaints of insidious onset, gradually progressive painless swelling on left side of the supraorbital region and proptosis, diplopia and lateral displacement of left eye since 18 months. Patient also had complaints of nasal obstruction and nasal discharge since 6 months.

Examination showed a smooth nontender, bony hard, nonmovable 3 × 3 cm swelling at the left supraorbital region. Proptosis was painless and gradual in onset. Eye was displaced laterally, inferiorly and outwards (Fig. 1). Nasal cavity was obliterated by a red pinkish mass covered by discharge pushing the septum to the opposite side. Nasal mass did not bleed on touch. It seemed to arise from the upper part of nasal cavity. Vision was normal. Posterior rhinoscopy was normal. Assessment of all cranial nerves was normal. There was no clinical sign of neurofibromatosis.

CT scan showed an expansile mass involving right frontal sinus, anterior and posterior ethmoid cells, left nasal cavity and superomedial compartment of left orbit with



Fig. 1: Clinical photograph of the patient showing frontal swelling, proptosis and displaced eyeball

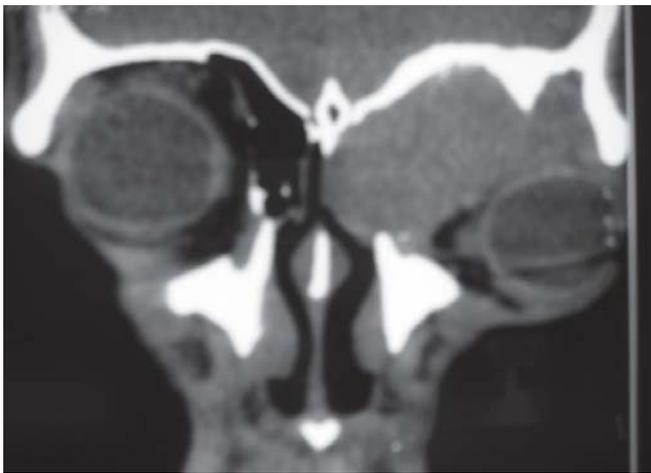


Fig. 2: CT scan picture showing homogenous soft tissue density in the supraorbital region displacing the eyeball

expansion and thinning of both tables of frontal bone (Fig. 2). Left eye ball was displaced anterolaterally. There was no evidence of bony erosion. Bony outline of paranasal sinus was intact. Tumor mass did not have any intracranial connection. On contrast administration mass showed mild enhancement.

Patient was operated under general anesthesia. An incision was made at the supraorbital rim just below the eyebrow extending from glabella to the lateral canthus of left eye. After opening the outer table of right frontal sinus which was grossly expanded and thinned out, we found a well encapsulated, firm mass that was filling the left frontal sinus and ethmoid region. Complete enblock excision of tumor was done. Inner table of frontal sinus and periorbital wall were intact but thinned out. There was no intracranial communication. The surgical specimen was $6 \times 3 \times 3$ cm, lobular, pinkish gray in color and firm in consistency. Cut surface was solid. Histopathology showed indistinct cytoplasmic boundaries with eosinophilic cytoplasm. Tumor cells arranged in whorled pattern with several psammoma bodies.

Immunohistochemistry study were performed in paraffin embedded tissue. The tumor cells had strong reactivity with cytokeratin and epithelial membrane antigen (EMA).⁹ The tumor cell did not stain with CD21 and CD23 antigen. Postoperative CT scan showed no residual mass. Patient is being followed up till date with no recurrence.

DISCUSSION

The intradiploic meningioma of paranasal sinuses is an extremely rare variety of ectopic meningioma. The origin of that tumor is thought to be from displaced meningeal

tissue in skin and subcutaneous tissue during embryogenesis or arise from ectopic arachnoidal cells within cranial nerve sheath of face and neck.¹⁰ Meningioma suppressor gene is located on the long arm of chromosome 22. Cytogenetic investigation of meningioma has revealed deletion of the long arm of chromosome 22.¹⁴ The extracranial meningiomas can be divided in four groups according to the etiology of development.¹¹

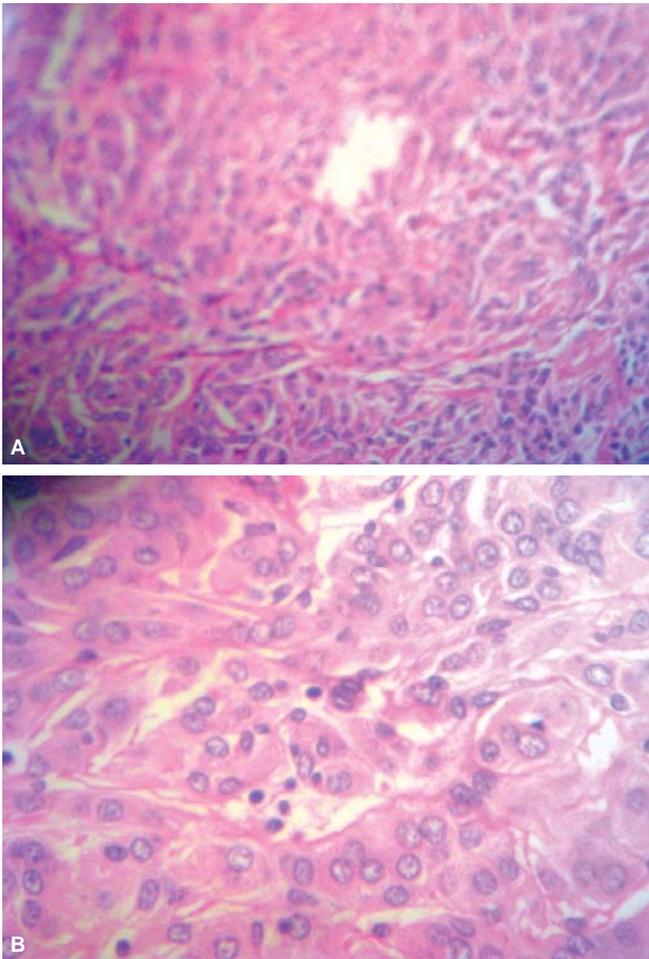
1. Extracranial extension of a meningioma with an intracranial origin (secondary).
2. Extracranial metastasis from an intracranial meningioma.
3. Extracranial extension of a meningioma arising in a neural foramina (primary).
4. Ectopic without any connection either with any foramina or any cranial nerve or intracranial structure (primary).

The present case is a ectopic meningioma of fronto-ethmoidal sinus. The tumor does not have any intracranial communication either clinically or radiologically.

Meningiomas are slow growing nonmetastatic benign tumors. They can permeate through cavities, foramina and cause pressure necrosis. So meningiomas usually produce pressure symptoms. The present case also had complaints of slow growing swelling and proptosis since 1.5 years with no involvement of orbit or any cranial nerves.

Due to many clinical, topographical, radiological and surgical factors, the history is not solely decisive for the prognosis of meningiomas.¹³ Imaging studies should be done in these cases. CT scan is confirmatory for extracranial primary meningiomas. Ectopic meningiomas should not have any intracranial extension or underlying bone erosion of the skull base which can be detected by the CT scan.

CT scan of the present case also showed an enhancing mass of left paranasal sinus, causing thinning of both tables of frontal bone, with no intracranial extension or any bony erosion of skull base. Final diagnosis is made by the histopathological examination but sometimes histopathological examination may be inconclusive.¹² Immunohistochemical study should be performed for confirmation. The microscopy of meningioma showed round, oval shaped cells with a pale eosinophilic cytoplasm, the cytoplasmic borders were indistinct. The tumor cells were arranged in nests of cells with whorled pattern. Psammoma bodies are also found in the tumor tissue. Cellular atypia and mitotic activity is not found.¹⁵ Tumor cells exhibit intense reactivity for vimentin and epithelial membrane antigen (EMA). The neoplastic cells did not stain with myelin basic protein, neurofilament protein, glial fibrillary acidic protein CD21 and CD23.¹⁶ In the present case, tumor cells showed lobular microarchitecture and indistinct cytoplasmic boundaries with whorled appearance on hematoxylin and eosin staining



Figs 3A and B: HPE picture showing tumor cells arranged in whorled pattern with psammoma bodies

(Fig. 3A). Tumor tissue also showed several psammoma bodies (Fig. 3B).

Primary surgical excision is the treatment modality of choice. Postoperative radiotherapy is not required. Complete enblock surgical excision was done in the present case because of the well-encapsulated nature of the tumor which is present in a bony shell. Chance of recurrence may be higher in the cases of atypical and malignant histological types, incomplete surgical removal and presence of mitosis on histopathological examination. Postoperative radiotherapy may be given in the malignant variety of meningioma or unresectable tumors.

CONCLUSION

Intradiploic meningioma is extremely rare benign tumor of the paranasal sinuses. It is often confused with other nasal masses and final diagnosis depends on histopathological examination and immunohistochemistry. Imaging studies should be done preoperatively. Complete surgical excision is the treatment modality of choice.

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