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CASE REPORT

A primary extracranial meningioma presenting as a painless nasal root mass

Sarah Hassan[1], Daniela Bondin[2] ,Emma Kitlowski[2] and Neel Umapathy[2]

[1]*Colorectal department,* [2]*ENT department, Royal Bolton Hospital, Bolton, England*

ABSTRACT

Extracranial meningioma remains a rare entity with few cases reported in literature. This report illustrates a unique case of a 28-year-old woman presenting with a deformity at the root of the nose which was investigated and treated as a primary extracranial meningioma. We discuss the presentation of this rare lesion and the challenges in its diagnosis and management. Literature has been reviewed to provide guidance for clinicians in diagnostic methods. A 28-year-old caucasian woman presented to the ear, nose and throat (ENT) clinic with a painless swelling at the root of the nose. Fine needle aspirations identified cells of an uncertain nature therefore an open biopsy was undertaken. This revealed features in keeping with a benign meningioma. Contrast MRI scan revealed an enhancing lesion involving the crista galli and cranial aspect of the nasal bone. No parenchymal involvement was found and the tumour was categorised as a primary extracranial meningioma. Multidisciplinary involvement from maxillofacial, neurosurgeons and ENT surgeons was required to arrange suitable surgical excision due to local extension of the tumour. The patient was warned of serious side effects including the risk of complete anosmia.

Key words: extracranial, meningioma, primary extracranial meningioma.

INTRODUCTION

Extracranial meningiomas are rare occurrences which constitute less than 2% meningiomas.[1] A large proportion of these are secondary lesions with a primary intracranial component.[2] The condition generally has a good prognosis and treatment of choice is surgical excision. Prognosis is poorer in disease recurrence however this is often due to incomplete excision rather than true recurrence.[2] This report describes an unusual case of a primary extracranial meningioma in a young patient presenting with a painless lump over the bridge of the nose.

CASE REPORT

A 28-year-old woman presented to the ear, nose and throat clinic with a 12 month history of a painless lump over the bridge of the nose. It had gradually increased in size with four self limiting episodes where the overlying skin became inflamed. She was otherwise well with no significant family history. She smoked twenty cigarettes per day. On examination there was a firm, non tender mass at the root of the nose, slightly larger on the left side.

Flexible nasoendoscopy noted a small septal deflection toward the left side.

The patient was reviewed 6 weeks later following a CT which showed clear sinuses and osteomeatal units and no osseus lesion (Fig. 1).

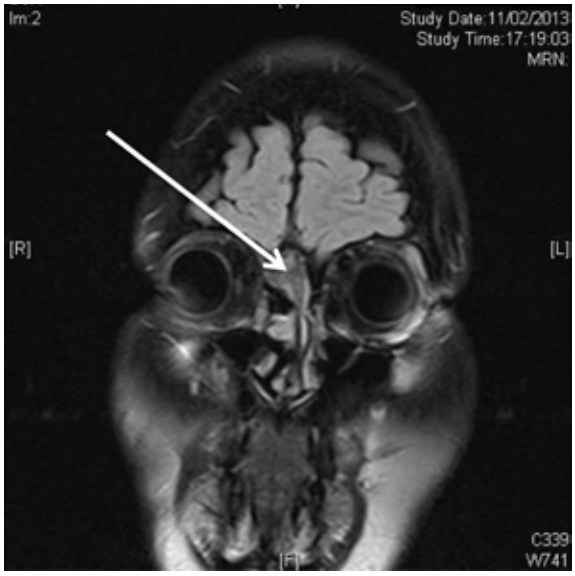

Figure 1, Craniofacial Contrast MRI which revealed an ehancing lesion involving the crista galli and cranial aspect of the nasal bone with an enhancing dural tail on the right extending between frontal lobes with no parenchymal involvement.

At this stage she reported that the lump had increased in size. Two fine needle aspirations were sent which identified cells of an uncertain origin. An open biopsy was requested by pathology for further information. A craniofacial contrast MRI was also requested. This revealed an enhancing lesion involving the crista galli and cranial aspect of the nasal bone. The lesion appe-

Correspondence:
Sarah Hassan
Colorectal Department, Royal Bolton Hospital, Bolton, England.
Email:sah87@doctors.net.uk

ared confined to bone at that stage but a subsequent MRI eight months later revealed an enhancing dural tail on the right, extending between the frontal lobes in the inter hemispheric fissure with no parenchymal involvement (Fig. 2).

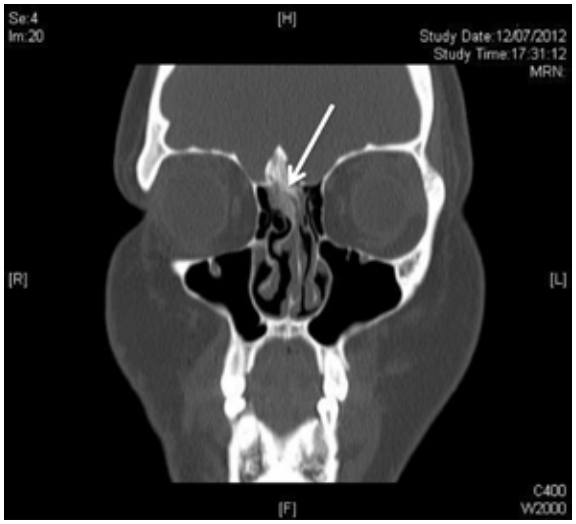

Figure 2, CT which revealed clear sinuses and osteomeatal units and no osseus lesion

The biopsy sample contained adipose tissue and fibrosis suggesting an infiltrating soft tissue lesion. There were scattered islands of cells with uniform round to oval nuclei and indistinct cytoplasm. Focally the cells formed whorls. No necrosis, cell atypia or mitotic figures were idenitifed. Immunostaining for vascular markers were negative. The lesional cells were focally positive for S-100 and Epithelial Membrane Antigen. The sample was reviewed by Christies Hospital, Manchester, who felt that features were in keeping with a meningioma; either an ectopic soft tissue meningioma with cutaneous extension or extension of a primary intracranial meningioma. As there were no atypical or malignant features it was classified a WHO grade I meningioma. The patient was referred to neurosurgeons at Hope hospital and following skull base multidisciplinary team meeting the decision was made for surgical excision. At this stage it is being managed as a primary extracranial meningioma.

Although the lesion is most likely benign it is not without its potential complications which are mainly due to local extension. It has been explained to the patient that the tumour extends into the cribiform plate therefore surgery will most likely compromise both olfactory nerves which could lead to complete anosmia.

DISCUSSION

Extracranial meningiomas remain rare with few cases reported in literature. They occur most commonly in females aged 45.[3] They account for less than 2% all meningiomas,[1] are theoretically derived from ectopic arachnoid cells[2] and classified according to the presence or absence of intracranial attachments. While primary extracranial meningiomas have no evidence of direct attachment to the brain, the majority of extracranial lesions are a secondary location of a primary intracranial tumour.[2] It is therefore imperative to investigate this once a diagnosis of meningioma has been made. The most frequent extracranial sites encountered are the nasal cavity and paranasal sinuses, cranial bones, middle ear, scalp and soft tissues of the face and neck and parathyroid gland.[1,2] There is only one report of a primary extracranial meningioma which has originated from the nasal septum thus far.[1]

The tumour remains a challenging diagnosis because growth rates are usually slow and most patients present with an asymptomatic disfiguring swelling which could be clinically correlated to other conditions, for example, Osteoma, Pagets disease, Fibrous dysplasia or a metastatic lesion. Radiological imaging can assist in diagnosis however does not necessarily provide a definitive diagnosis for the nature of the lesion. Therefore histology remains gold standard. This is demonstrated in this case. Important CT and MRI features include the appearance of calcification, intracranial extension and marked contrast enhancement with a typical dural tail.[4] This was only identified on the second MRI in this case.

Further tests to confirm diagnosis of a meningioma include immunohistochemical staining.[3] Meningiomas have high positivity testing for vimentin and epithelial membrane antigen, and a weaker positivity testing for S-100 protein.[1,2,3] This correlates with the findings in this case where the specimen was positive for epithelial membrane antigen and S-100 protein.

In terms of prognosis, WHO classification 2007 categorises extracranial meningiomas according to their histological appearance and clinical behaviour. WHO grade I tumours generally have an excellent prognosis with infrequent recurrence rates. They are benign with transitional, psammomatous, fibrous, meningothelial, angiomatous or secretory histological patterns. Malignant meningiomas (WHO grade III) are usually fatal with higher rates of recurrence and metastasis. Studies have shown a 5 years mortality rate of 68%.[3]

Current treatment of choice for a localised primary extracranial meningioma is surgical excision with no documented beneficial evidence of radiotherapy or chemotherapy, particularly in benign disease.[4,5] It is important to consider the location of the tumour and potential disastrous effects to local surrounding structures as discussed in this case. Imaging, tissue biopsy and multidisciplinary team involvement are invaluable in diagnosis and management of this rare lesion.

CONCLUSION

To summarise, primary extracranial meningioma is a rare condition which often presents in a transient manner making diagnosis quite difficult. There are diagnostic features in imaging but essentially a histological diagnosis is key.

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