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Case Presentation

Aggressive meningioma presents as innocuous forehead lesion: a case report

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Abstract

Cutaneous meningiomas are very rare neoplasms. In this case report we document a type III (anaplastic meningioma) presenting as a subcutaneous forehead mass. Anaplastic meningiomas arise from the neuraxis. They are biologically aggressive neoplasms that extend into the dermis or subcutaneous tissue via direct extension through the bone.

Keywords: Dermatology, meningioma, forehead, lesion, cutaneous meningioma, osteolytic meningioma, epithelioid, craniectomy

Introduction

Meningiomas are common neoplasms arising in the neuraxis. Asymptomatic meningiomas are discovered in 1-2% of people as incidental findings at autopsy. In addition, they are frequently discovered as incidental findings with brain imaging procedures. More than 90% of meningiomas are benign (grade I), 7% are atypical (grade II), and 2% are anaplastic (grade III). Risk factors for meningioma are brain radiation, brain injury, neurofibromatosis type 2 (NF-2), female gender, and increasing age.

"Cutaneous meningiomas" are very rare neoplasms with both congenital and acquired forms. Type I cutaneous meningiomas are present at birth on the scalp or paravertebral areas. These tumors develop from ectopic arachnoid cells trapped in the dermis and subcutaneous tissue during development owing to failure of neural tube closure. Type II cutaneous meningiomas extend to the skin by contiguity around the eyes, ears, nose, and mouth. In this condition, there is no subjacent meningioma of the neuraxis. It has been suggested that these neoplasms are formed by remnants of arachnoid cells extending along cutaneous nerves. Type III cutaneous meningiomas extend into the dermis or subcutaneous tissue by direct extension through the bone or surgical or traumatic defects in the bone from a neoplasm arising in the neuraxis. In a series of 92 cases of cutaneous meningioma, only 4 were classified as type III neoplasms. The most common site for type III meningioma is in the occipital area [21].

We report a single case of a type III meningioma presenting as a subcutaneous mass on the forehead.

Case synopsis

A 76-year old man presented for his annual primary care visit for follow up of well-controlled hypertension with a new "slightly painful" growth on the forehead for several weeks. The lesion was described as "not indurated" and 4 cm in diameter (Figure 1). The working diagnosis was subfrontalis lipoma. The patient was referred to a general surgeon for treatment. Instead of a lipoma,

the surgeon encountered friable tissue and a frontal bone defect. A computerized tomographic scan of the head revealed a large mass centered within the anterior cranial fossa. The lesion extended through the right frontal bone (Figure 2).

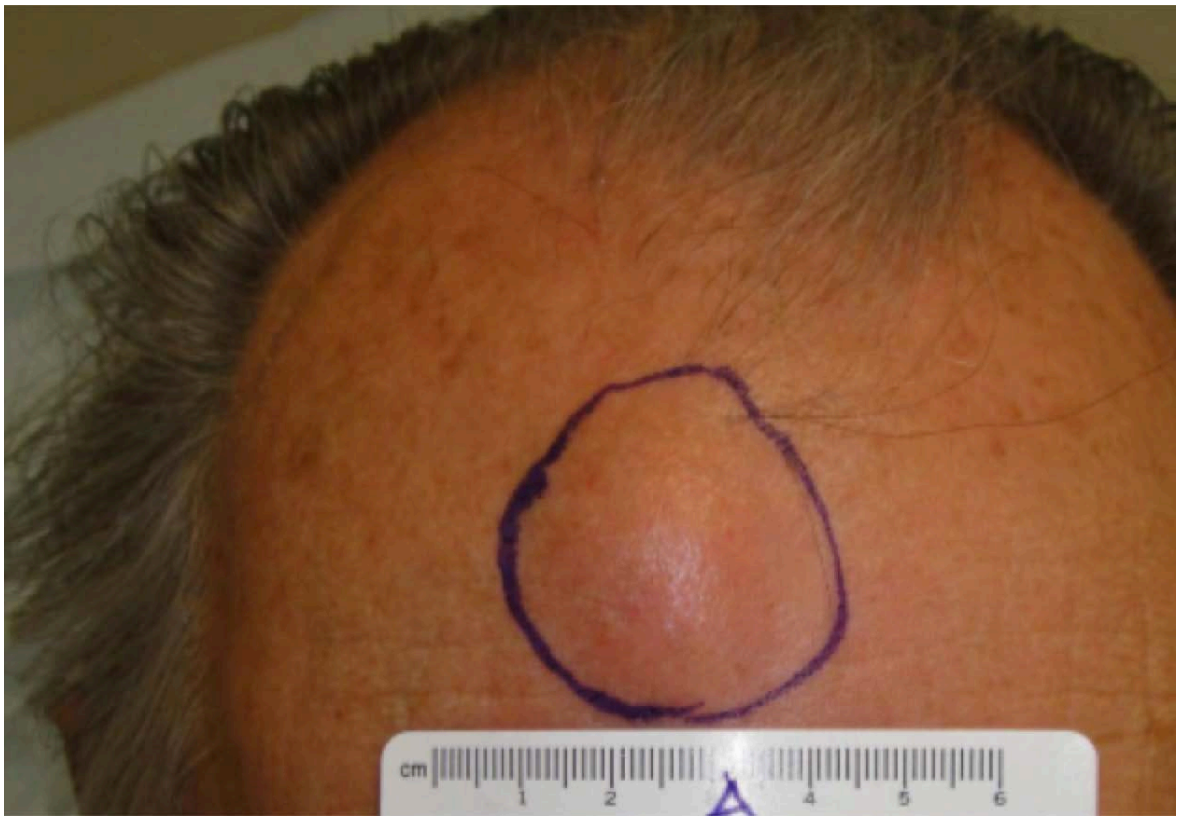

Figure 1. Photograph of the lesion reveling a mass 4cm in diameter

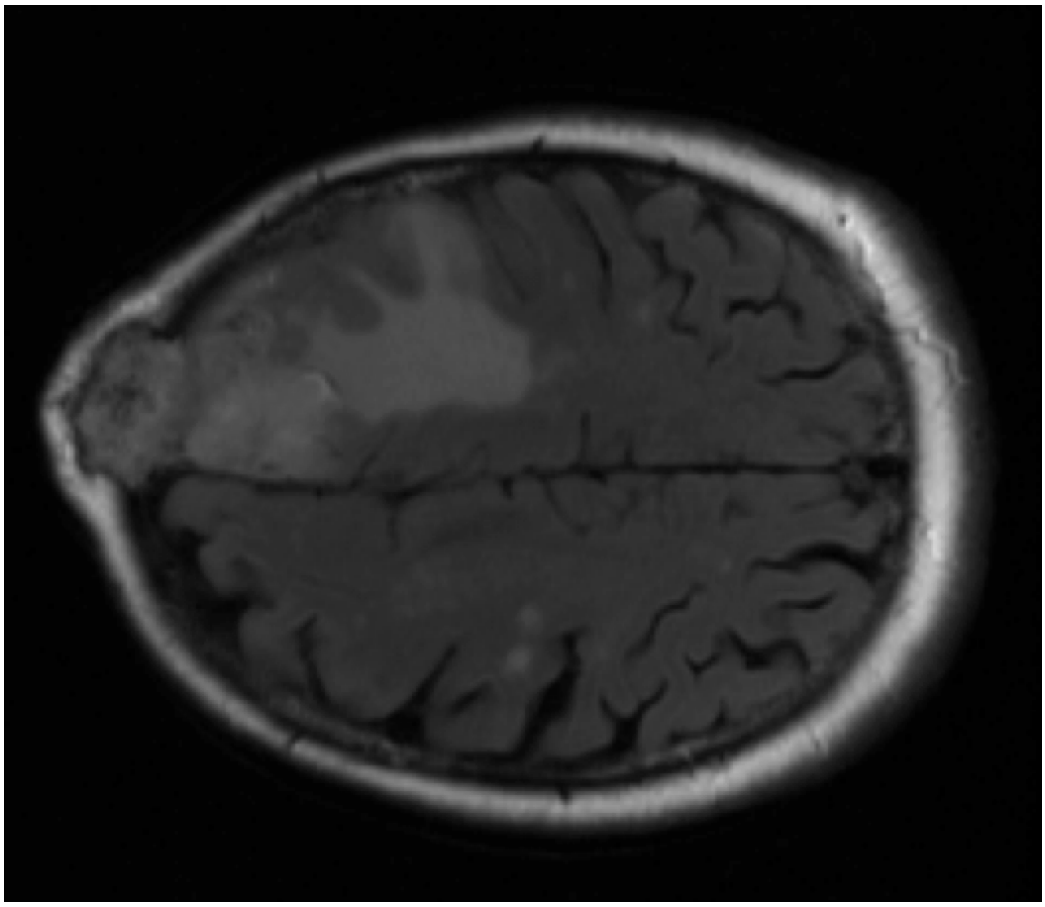

Figure 2. Head CT showing a large mass within the anterior cranial fossa, extending through the right frontal bone

Microscopic examination of the tissue revealed a poorly differentiated epithelioid neoplasm with secretory features (Figure 3). The neoplasm showed reactivity with pancytokeratin, vimentin, and focally with cytokeratin-7, cytokeratin 5/6, p63, and progesterone receptor. It did not show reactivity with antibodies to S-100, HMB-45, transthyretin factor 1, prostate specific antigen, prostate specific acid phosphatase, chromogranin, or synaptophysin. Ki-67 showed a high proliferative index (~75%). Seventeen mitoses were counted in 10 high power fields. PAS special staining shows abundant secretory globules (pseudo psammoma bodies).

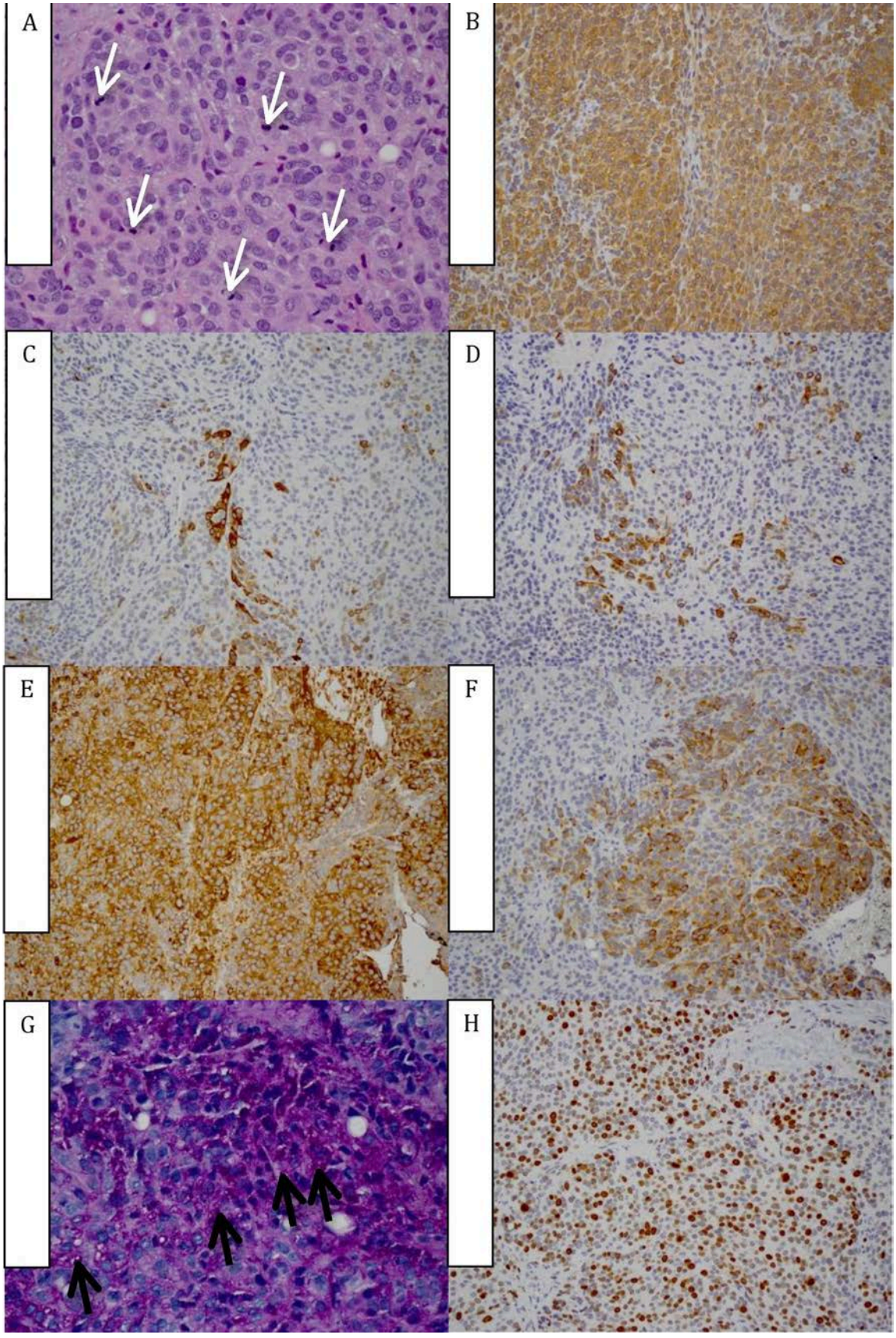

Figure 3. Secretory meningioma. Routine, hematoxylin and eosin, stained slide showing an atypical epithelioid neoplasm (A) with mitotic activity (white arrows). Diffuse immunoreactivity to pankeratin (B) and vimentin (E) is noted, but only patchy reactivity to Cytokeratin 5/6

(C), Cytokeratin 7 (D), and Progesterone receptor (F) is seen. Pseudopsamomma bodies are identifiable on PAS staining (black arrows) (G). Ki-67 shows a high proliferative index (~75%)(H).

A bifrontal craniectomy was performed with resection of the tumor followed by a cranioplasty. No brain invasion was discovered. The surgical wound healed well with no residual neurologic deficits. The patient remained well and asymptomatic for six months, when he developed three new growths on the scalp. Magnetic resonance imaging revealed masses with both intracranial and extracranial components consistent with recurrent meningioma. He subsequently developed severe back pain; metastatic disease was discovered. Palliative radiation was administered. He died 10 months after diagnosis.

Discussion

Cutaneous type III meningiomas are uncommon biologically aggressive neoplasms. Miedema and Zedek [1] reviewed and summarized the literature. The mean age of patients was 54 years. Most of the lesions occurred around the face, temple, and scalp and presented as slowly growing subcutaneous masses. Although meningiomas are more common among women (2:1 ratio), atypical or anaplastic meningiomas are reported more in men [35]. Our patient presented with an extracranial subcutaneous mass extending from an intracranial osteolytic meningioma.

The histologic features typically show atypical epithelioid cells. Immunohistochemical studies are useful to rule out neoplasms of vascular, smooth muscle, histocytic, or melanocytic origin. Positive reactivity with antibodies to pankeratin, vimentin, and epithelial membrane antigen are consistent with a meningioma diagnosis. Our case showed additional patchy reactivity with cytokeratin 7 and cytokeratin 5/6; abundant secretory globules by PAS special staining were identified, consistent with secretory meningioma.

Although rare, cutaneous meningiomas should be considered in the differential diagnosis of subcutaneous nodules presenting on the head and neck particularly when accompanied by neurologic signs and symptoms [21]. Without neurologic signs and symptoms to direct brain imaging studies, incisional biopsy is the only reasonable way in which this diagnosis could be made, yet biopsy may be associated with increased risk for cerebral infection, CSF loss, and worsening neurologic symptoms [6]. When this diagnosis is suspected on the basis of neurological signs and symptoms accompanied by brain imaging studies, Kalfa [24] has suggested fine-needle aspiration as a minimally invasive method to obtain a tissue diagnosis.

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