Case Report

Intraosseous Angiolipoma of the Cranium: Case report and review of the literature

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Abstract

Background: Angiolipomas are benign tumors that usually occur in patients during their late teens or early twenties. Most are found as multiple lesions and often located on the arm or trunk. Although predominantly subcutaneous lesions, intraosseous angiolipomas are primarily found in the mandible and ribs, with one case involving the calvarium reported in the literature.

Case Description: We report the case of a 55-year-old male who presented with headache, nausea, vomiting, and double vision and was found to have an invasive ductal breast carcinoma. The patient subsequently underwent a resection and the lesion was found to be an angiolipoma.

Conclusion: We report a rare case of an intraosseous angiolipoma in the setting of a male with invasive ductal breast carcinoma and a review of the literature.

Key Words: Angiolipoma, calvarium, intraosseous, invasive ductal breast carcinoma

INTRODUCTION

Angiolipomas are benign tumors that usually occur in patients during their late teens or early twenties. Most are found as multiple lesions and are often located on the arm or trunk, with the forearm being the most common site. Although predominantly subcutaneous lesions, intraosseous angiolipomas have a predilection for the mandible and ribs, with one case involving the calvarium reported in the literature. Herein, we report a rare case of an intraosseous angiolipoma of the calvarium found in a male patient who was concurrently diagnosed with invasive ductal breast carcinoma.

CASE REPORT

A 55-year-old right handed male with a past medical history of diabetes, hypertension, and a greater than 10 pack-year history of cigarette smoking, presented with headache, nausea, vomiting, and worsening double vision. On physical examination, the patient was alert and oriented. His cranial nerves were intact. He was noted to have facial asymmetry and slight mass over the right forehead. Further on examination it was noted that he had a lesion on his right breast and nipple that he reported had been present for approximately one year, with occasional enlargement, bleeding, and purulent...
discharge. He had no lymphadenopathy on examination. Noncontrast computed tomography (CT) head indicated a 4.3 × 2.2 × 1.7 cm right frontal calvarial lesion, with internal trabeculated bone mixed with fatty components, and no associated involvement of overlying soft tissue or brain parenchyma [Figure 1]. Magnetic resonance imaging (MRI) of the brain confirmed the presence of a right sided heterogeneously enhancing calvarial lesion with mild mass effect on the adjacent frontal lobe [Figure 1]. A CT of the chest, abdomen, and pelvis showed a soft tissue density in the right retroareolar space, corresponding to the location of the painful breast lesion, and associated with enlarged right hilar and subcarinal lymph nodes.

The patient initially underwent a punch biopsy of the breast lesion and subsequent histopathologic analysis revealed the lesion to be invasive ductal carcinoma. In order to obtain a tissue diagnosis and relieve symptoms caused by the calvarial lesion, the patient underwent a right frontal image-guided craniectomy. A modified bicoronal incision was carried out with exposure of the mass. Using image guidance, a craniectomy was carried out with en-bloc calvarial tumor resection. There was no dural involvement. A titanium mesh cranioplasty was performed. Postoperative imaging showed no residual tumor and the patient was discharged on postoperative day 2 with improvement in his headaches. The patient later underwent a modified radical mastectomy and biopsy of an axillary sentinel node. Pathology and staging demonstrated a T4aN1aM0 Stage IIIb infiltrating ductal carcinoma.

The pathologic calvarial specimen consisted of a 5.5 × 5.5 × 0.8 cm portion of calvarium involved by an expansile lesion measuring 3.5 × 3.5 × 1.5 cm. The lesion was well-circumscribed and showed mixed areas of firm-to-soft tan and red lobulated tissue. Histopathologic analysis performed on the specimen proved the tumor to be an angiolipoma. Microscopically, the lesion consisted of mature trabecular bone intermixed with mature adipocytes and variably sized dilated vascular channels [Figure 2]. The adipose component was mature and composed of normal appearing lobules separated by thin, fibrous septa. No cytological atypia, hyperchromasia, mitoses, or lipoblasts were present. No increased cellularity or epithelioid cells were noted, and immunohistochemical stains for pankeratin were negative.

DISCUSSION

The case described herein involves a male patient presenting with an intraosseous angiolipoma involving the calvarium, and concurrent metastatic ductal carcinoma of the breast. Initially, the authors were suspicious that the lesion represented a metastasis from the breast. Intraosseous calvarial metastases from breast cancer occur in approximately 3% of patients. Angiolipomas are benign lesions that generally occur in the subcutaneous tissues. Only seven instances of intraosseous angiolipoma have been reported in the literature and, of these, only one involving the cranium. Yu and colleagues described the case of a 39-year-old male who presented with right parietal swelling and, upon further evaluation, was noted to have a nonenhancing mass within the calvarium. Though he remained neurologically asymptomatic, the patient eventually underwent a

Figure 1: Noncontrast CT head axial images indicate a right frontal bony lesion with mixed fat and boney trabeculae. (a) Brain windowing, (d) Bone windowing. MRI brain, (b) T1-weighted image, (c) Contrast enhanced image, (e) Sagittal T1 image, (f) Coronal contrast enhanced image.
two-staged surgical resection after the lesion grew to being 7 cm in size. Complete excision of the tumor was accomplished, and the patient did not require any further treatment.

Radiographically, intraosseous angiolipomas are seen to arise within the marrow space and expand the bone. Typically, on CT imaging, the lesion exhibits characteristics consistent with its fatty component, in addition to having bony trabeculae and septations. Intraosseous angiolipomas generally appear hyperintense on T1 MRI, with enhancement and T2 imaging demonstrating flow-voids, indicating their significant vascularity.

Histologically, intraosseous angiolipomas are composed of mature fat and vessels, without lipoblasts, atypia, or an epithelioid component. While interior vessels tend to be of small caliber, those located at the periphery may be quite prominent and give the impression of vascular malformation. A characteristic finding, which helps to distinguish angiolipoma from the more common lipoma, is the presence of scattered fibrin thrombi, which was seen in our specimen. Mast cells may also be present, another feature, which may assist in distinction from lipoma. Infiltrating angiolipoma, an unencapsulated variant that often invades bone and adjacent tissues, is known to be more aggressive clinically.\(^\text{1,2}\) Although still considered benign, complete surgical resection of this entity is more challenging.

Genetic causes for these tumors have been evaluated in familial syndromes such as familial angiolipomatosis, which is a benign autosomal-dominant condition. This results from a translocation of genes on chromosomes 12 and 3. To date, genetic testing on spontaneous angiolipoma’s have been of little yield.\(^\text{1,3}\)

CONCLUSION

We present a rare instance of a solitary cranial intraosseous angiolipoma in a patient with concomitant metastatic ductal carcinoma of the breast. Given the patient’s clinical presentation and radiologic findings, diagnosis of osseous metastasis to the calvarium was initially favored. However, following operative en-bloc resection, the lesion exhibited characteristic histologic findings of an angiolipoma. This case represents only the second report of a calvarial angiolipoma in the extant literature.

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Commentary

The authors report a rare case of a calvarial osseous lesion in the setting of a male patient with invasive ductal breast carcinoma. The calvarial lesion was resected to alleviate symptoms of raised intracranial pressure, with histology revealing an angiolipoma. The authors report a peculiar event where two unrelated extremely rare tumors have been identified concomitantly. The incidence of male breast cancer in the general US population is <1%. Angiolipoma is a benign tumor generally found in subcutaneous tissue, with one other cranial intraosseous lesion reported in literature. The authors initially favored a diagnosis of metastasis to the calvarium, but following an excision biopsy, turned out to be an intraosseous angiolipoma. As physicians, we are trained to utilize the principles of diagnostic parsimony. When faced with clinical dilemmas in patients with multiple problems, the medical adage “when you hear hoof beats behind you, think horses, not zebras”, comes to mind.

A few years ago I had the opportunity to partake in the treatment of a 50-year-old female patient presenting with an acute history of raised intracranial pressure, in the absence of any neurological deficit. Her magnetic resonance imaging (MRI) revealed a small left intraventricular lesion (measuring 1.7 × 1.7 cm) without hydrocephalus, a computed tomography (CT) scan of her chest, abdomen, and pelvis revealed a concomitant renal cell mass. On imaging, the intraventricular mass appeared characteristic of a left atrial meningioma. Faced with the uncertainty of a clear diagnosis, a whole body (18) F-fluoro-2-deoxy-d-glucose positron emission tomography scan (FDG-PET) and an Indium¹¹¹-octreotide brain scintigraphy (OBS) imaging were performed. The ability to differentiate a benign (meningioma) from a malignant (renal cell carcinoma) metastatic lesion using the information obtained would help decide a management strategy. The FDG-PET and OBS imaging was discordant for these lesions, the renal mass was positive (increased uptake) on the FDG-PET and negative (absence of tracer uptake) on OBS study. The intraventricular lesion was positive on the OBS study and negative on the FDG-PET scan. She underwent a nephrectomy with a diagnosis of renal cell carcinoma. The imaging results favored an intraventricular meningioma, the small tumor size and absence of hydrocephalus made Gamma Knife Radiosurgery (GKRS) with 15 Gy to the 51 isodose line (IDL), the initial treatment of choice. About 14 months later, she presented with recurrent symptoms of raised intracranial pressure, her MRI revealing an increase in the size of the intraventricular lesion along with hydrocephalus. The tumor was now amenable to surgery and she underwent a left temporal lesionectomy, histopathology confirming a renal cell carcinoma metastatic lesion. She underwent GKRS to the resection cavity bed, receiving 15 Gy to the 51 IDL and without any subsequent intraventricular recurrence.

Metastatic lesions from renal cell carcinoma have a propensity to present as late as 10 years following diagnosis of the primary tumor, even though they are the fourth most common to metastasize to the brain. The combined use of noninvasive imaging modalities, OBS, and FDG-PET that are presently available have been reported to significantly increase diagnostic sensitivity and specificity for intracranial meningiomas. These tests have enabled differentiation of benign from malignant tumors, and have been used in the presence of multiple tumors, to identify tumor grade and possibly providing evidence suggesting a collision tumor. The high index of suspicion for a particular diagnosis may be reenforced with the use of noninvasive testing, especially in circumstances where observation, stereotactic radiosurgery, or alternative medical treatment options are available.

The authors’ in their case report conclude that were it not for a histological diagnosis, they would have assumed the calvarial lesions to be metastatic. In our case, the evidence favored a benign intraventricular lesion but final histopathology revealed it to be metastatic. Modern imaging techniques provide a basis for narrowing the differential diagnosis but none of the modalities can presently be considered to be sensitive and specific enough to replace a tissue diagnosis. Although surgical interventions ranging from diagnostic stereotactic biopsy to open excision biopsy is associated with risks, histopathological diagnosis remains the gold standard.
The patient should be made aware of the risk-benefit ratio of their investigations, interventions, and play a part in deciding the most suitable management options available to them. This case report indirectly emphasizes the importance of a judicious tissue diagnosis from intracranial lesions.

We always strive to avoid harm to our patients and aim to provide them with safe, cost effective, and beneficial treatments. As treating surgeons, we face challenging situations in evaluating patients’ with complex and multiple medical problems, so when we ‘hear hoof beats, we have to think of both horses and zebras’. Herein lies the importance of experience and clinical judgment, enabling us to decide how and when to use laboratory tests, imaging, and appropriately time surgical interventions.[1] To quote from Sir William Osler “The value of experience is not in seeing much, but in seeing wisely”.

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