Case Report

International Journal of Neurology and Neurosurgery
Volume 9 Number 2, July - December 2017
DOI: http://dx.doi.org/10.21088/ijnns.0975.0223.9217.17

# Giant Intraosseous Angiolipoma of Skull: A Case Report and Review of the Literature

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**Abstract**

Intraosseous angiolipomas are very rare tumors occurring most commonly in the ribs and mandible. So far, only five cases of intraosseous angiolipomas of skull have been reported in the literature. We report the rare case of a 52 years old lady admitted with a swelling over the right frontotemporoparietal region of 12 years duration, which turned out to be an angiolipoma. We discuss its radiological features, histological features and management. We also present a review of the literature on intraosseus calvarial angiolipomas.

**Keywords**: Angiolipoma; Intraosseous; Skull.

## Introduction

Although angiolipomas can occur anywhere in the body, with the subcutaneous tissues of the trunk and upper arms being the most common sites, tumours involving the craniospinal axis are uncommon and almost always occur in the epidural space. Intraosseous angiolipomas are extremely rare, slow-growing tumors occurring most commonly in the ribs and mandible [6,7]. They are composed of mature adipose tissue admixed with arteries, veins, sinusoids and capillaries [1,6]. We report the case of a 52 years old lady admitted with a swelling over the right frontotemporoparietal region for 12 years which turned out to be an angiolipoma.

## Case Report

A 52 years old lady was admitted with complaints of sudden loss of consciousness from which she recovered spontaneously and then continued to have headache. Patient had a swelling over the right frontotemporoparietal region for 12 years. It was insidious in onset and had gradually increased in size. There was no pain over the swelling and patient had no history of trauma and headache in the past. On examination, the patient was conscious and oriented. There was no papillodema and cranial nerve deficit. She had left hemiparesis of power 4/5. She did not have sensory deficit. On local examination, there was a 10 * 8 cm size, hard, immobile mass in the right frontotemporoparietal region which was not pulsatile. The skin over the swelling was smooth and pinchable and there were no dilated veins over the swelling. There was no cough impulse.

The patient did not have any comorbid illness. The patient had consulted a private practitioner for the swelling and had refused surgery 8 years back.

The patient does not give history of similar illness in the family members.

*CT Scan of the Brain* (Figure 1) revealed mixed dense trabeculated intraosseous lesion in right frontotemporoparietal calvarium with intracranial expansion of the mass causing squashing of right lateral ventricle with minimal midline shift. Patient was initially treated with anticonvulsants and antioedematous drugs.

*MRI of the Brain* (Figure 2) revealed a lesion centered within the right frontotemporoparietal bone extending to the cranial vertex with corresponding calvarial expansion into the extra-axial space over

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**Received on** 13.04.2017, **Accepted on** 24.04.2017

the right frontal and temporal lobes with resultant cerebral compression. There was no radiographic evidence of osseous destruction or parenchymal edema. There were no other intra- or extra axial lesions identified.

The decision was made in conjunction with the patient's wishes to proceed for surgical resection.

Intra operatively, the protruding skull mass extended approximately 5 cm above the surface of surrounding bone and measured 8-10 cm in diameter. It was hard in consistency, greyish in colour and highly vascular. The tumor had infiltrated the dura, but it was not adherent to the arachnoid. Tumour was resected totally along with the infiltrated dura. Duroplasty was done with temporalis fascia. Three units of whole blood were transfused. Cranioplasty was deferred awaiting histopathological examination.

Gross cut section of the tumour (Figure 3) showed a trabeculated lesion with yellowish fatty tissue between the trabeculae.

*Histopathological Examination* (Figure 4) showed bony trabeculae with well circumscribed lobules of mature adipocytes intermixed with prominent, proliferating blood vessels. Thin fibrous septae were seen separating the adipocytes. No evidence of atypical or dysplastic cells was seen . Many of the blood vessels showed the presence of intraluminal thrombi. Some foci showed mast cells.

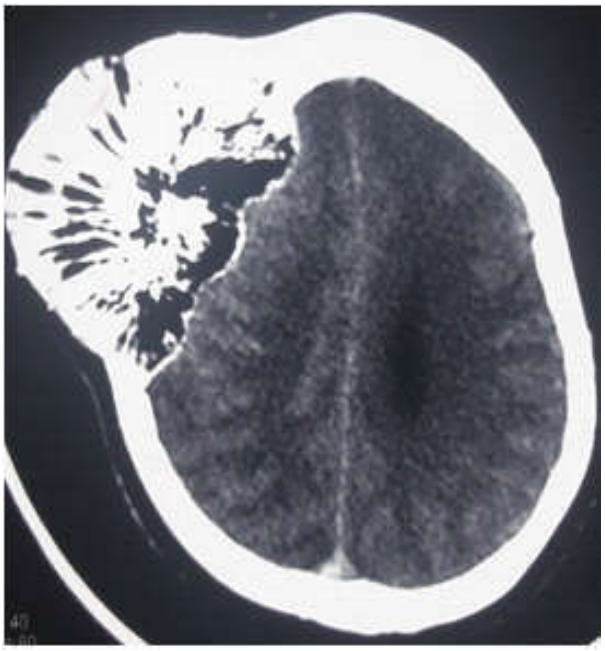

**Fig. 1:** *CT scan of the brain* revealed mixed dense trabeculated intraosseous lesion in right frontotemporoparietal calvarium with intracranial expansion of the mass causing squashing of right lateral ventricle with minimal midline shift.

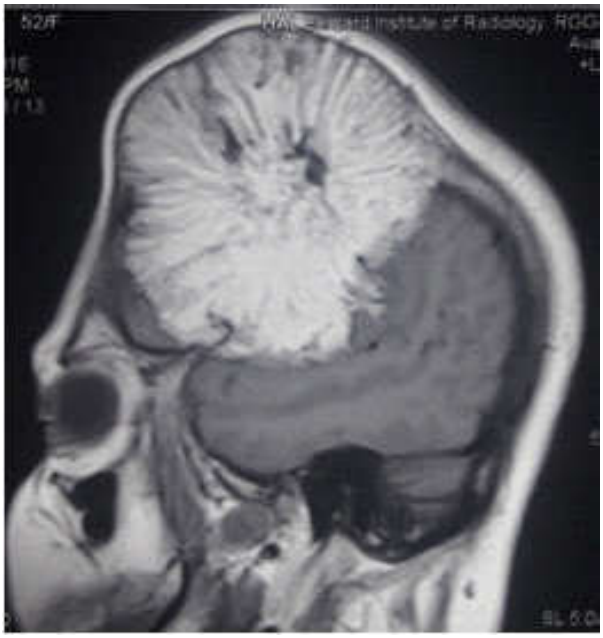

**Fig. 2: MRI of the brain** revealed a lesion centered within the right frontotemporoparietal bone extending to the cranial vertex with corresponding calvarial expansion into the extra-axial space over the right frontal and temporal lobes with resultant cerebral compression. There was no radiographic evidence of osseous destruction or parenchymal edema. There were no other intra- or extra axial lesions identified.

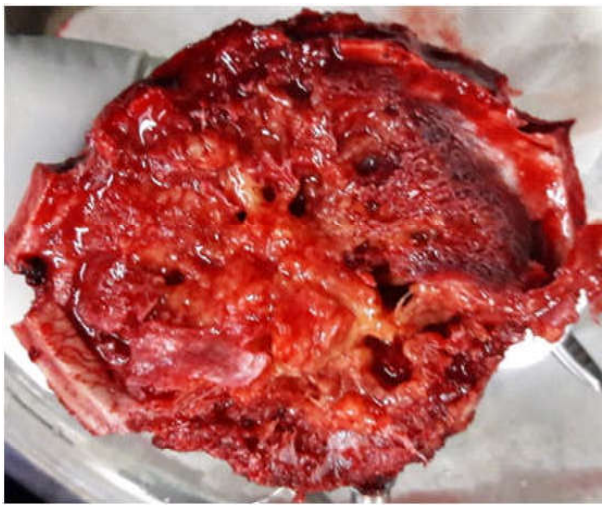

**Fig. 3:** Gross cut section shows a trabeculated lesion with yellowish fatty tissue between the trabeculae

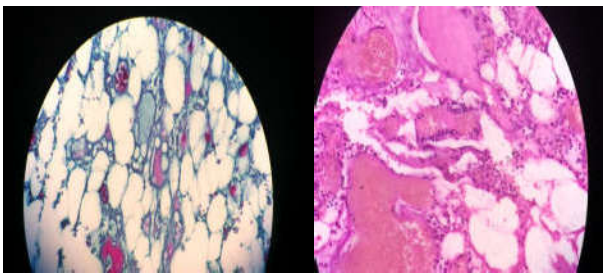

**Fig. 4:** Lobules of mature adipocytes with intervening proliferating blood vessels. Intraluminal fibrin thrombi – H & E and Masson's trichrome

Masson's trichrome stain confirmed the presence of fibrin thrombi.

Immunohistochemistry for epithelial membrane antigen (EMA) and cytokeratin was negative, ruling out an intraosseous meningioma. Hence a diagnosis of angiolipoma was made.

Post operatively patient developed a pseudomeningocele over the operative site. CSF Lumbar drainage was done for three days. Pseudomeningocele decreased in size and the remaining postoperative period was uneventful.

## Discussion

Intraosseous angiolipomas are extremely rare lesions mostly noted to involve extra-cranial locations such as the mandible and ribs. There are only five reported cases of intraosseous angiolipomas that involve the cranium, where they present as slow-growing masses.

Literature search was done on pubmed.org with the keywords angiolipoma, cranium, intraosseous, skull, bone. We have reported an intraosseous angiolipomas that involved the frontotemporo parietal region. The lesion presented as a hard, painless mass of 12 years duration, that is 10x8 cm in size, without causing significant bone erosion, but with expansion of calvarium and infiltration of the dura.

The symptoms of intraosseos angiolipomas vary depending on the location and size of the tumor. The most common first symptom is swelling, followed by hypoesthesia, and headache [4]. However, the tumor may remain asymptomatic for many years. Our patient presented to our hospital with loss of consciousness, head ache, vomiting and left hemiparesis.

The pathogenesis of angiolipomas is unclear. Most authors accept that angiolipomas may derive from embryonic sequestration of multipotential-mesenchymal cells and this process becomes activated at puberty by hormones. Some reports suggest that trauma is a causal agent. However, no history of trauma is seen in most angiolipomas. Similarly, there was no history of trauma in our case. In addition, fatty degeneration in hemangiomas and vascular proliferation in congenital lipomas have been implicated as possible predisposing factors. One of the hypotheses is for an association between mast cells and angiogenesis. It has been supposed that mast cells might play a role in the increased vascularity of angiolipoma. Shea and Prieto have reported angiolipomas to have 10 times the number of mast cells than classic lipomas (25.34 vs. 2.41/ $mm^2$). Mast cells strongly produce VEGF, which is known to be essential for angiogenesis, and TNF, which promotes inflammation [3]. Mast cells along with tryptase stimulate the proliferation of human vascular endothelial cells in human tumors. An average of 10 mast cells per 0.22 $mm^2$ (45 mast cells per $mm^2$) in (45 mast cells per $mm^2$) in Toluidine Blue stained slides was reported in one study. In our case, 20 mast cell per $mm^2$ were seen.

Angiolipomas are known to have a different morphology from other skull bone tumours such as lipomas, haemangioma, fibrous dysplasia,osteoma or meningioma. They are extremely rare, located in bones, and are composed of mature fat cells with numerous thin or thick walled small blood vessels. Vascular components may be patchy and predominantly capillaries. Angiolipomas have mature adipocytes, interspersed angiomatous proliferation, fibrin thrombi and presence of numerous mast cells.

There is absence of nuclear pleomorphism, mitoses, necrosis, and other mesenchymal elements (smooth muscle, neural tissue) in contradiction to other lipomatous lesions such as angiomyolipoma, angiofibrolipoma, angiomyxolipoma, liposarcoma, and hemangiomas. In the case of a hypovascular lesion, one possible differential diagnosis is lipoma. There is characteristic fibrin thrombi and presence of increased mast cells in an angiolipoma. The hypervascular lesion may be difficult to distinguish from hemangioma with entrapped fatty tissue and lipoma with prominent blood vessels. In hemangioma with entrapped fatty tissue, there is a little lipomatous tissue and no fibrin thrombi. Vascular lumens of conventional lipoma have no fibrin thrombi.

In our case, calcified, trabecular and expanded bone was present with marrow spaces occupied by small and large caliber blood vessels and adipose tissue. These vessels were separated by fibrous stroma. The vessel wall thickness was also variable and so was the size of the lumen. Fibrin thrombi were seen [3]. It may be hypothesized that the intra osseous angiolipoma (IOAL) may represent either hyperplasia of fat, with an associated increase in the vascular channels, or a true neoplasm.

The radiological appearance of angiolipoma resembles like that of a lipoma, hemangioma, Langerhans cell histiocytosis, epidermoid cyst, metastasis or fibrous dysplasia.

On CT imaging, IOAL appear as hypodense lesions that involve the full thickness of the cranium with

bony spicules. In our case, the lesion was mixed dense trabeculated intraosseous lesion.

On MRI brain, IOAL are of variablehyperintensity both in T1W and T2W images, with trabeculated pattern [1]. The heterogeneously increased signals on MRI indicate the presence of hemosiderin, methemoglobin, or oxy- and deoxy-hemoglobin within the diploic space.

The internal or external table is intact, but in some parts may be burst out by the tumor invasion. There is no remarkable enhancement after contrast material injection on T1W images. On FLAIR sequences, mixed hypo- and hyper-intensities of filliform pattern are compatible with mixed fatty and vascular tissue components. In our case, the lesion was hyperintense in T1 and T2, with no contrast enhancement. So, histopathological evaluation is mandatory for final diagnosis.

The treatment of choice is total excision which is curative.

## Conclusion

To the best of our knowledge, only five cases of Intraosseous angiolipomas of skull have been reported so far, with ours being the sixth. The diagnosis of angiolipoma of skull must be kept in mind in the preoperative differential diagnosis of these types of calvarial lesions. We have reported a unique case of an intraosseous angiolipomas involving the calvarium. Although fully benign in nature, intraosseous angiolipomas show a tendency to expand over years with potential for causing neurological symptoms, as in our case. Complete resection is usually curative.

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