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Intraosseous angiolipoma of the frontal bone with a unique location: A clinical and pathological case illustration and review of the literature

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Here, we report a case of a 16-year-old female patient was referred with scalp swelling and headache. Her neurological examination was normal and imaging of the skull revealed a well-defined lytic lesion measuring 15 mm x 6 mm to the right of the frontal bone. She was operated on with a prediagnosis of Langerhans cell histiocytosis. A wide excision with negative margins was made and the defect was reconstructed with a titanium plate. Subsequently, the lesion was histopathologically diagnosed as an angiolipoma of the frontal bone. The postoperative period was uneventful and she remained well during 1-year follow-up with no evidence of recurrence. Angiolipomas are rare benign lipomatous lesions located mostly in subcutaneous tissue of the forearm or trunk and frequently occur before puberty or in young adults. They are not common in bones. To the best of our knowledge, this is the first angiolipoma of the frontal bone reported.

INTRODUCTION

Angiolipomas are rare benign lipomatous lesions that differ from lipoma with their characteristic histology consisting of mature adipocytes and prominent, proliferated, thin walled blood vessels. [sup][1] They mostly occur before puberty or in young adults and commonly present as multiple, painful, slow growing, small nodules (<2 cm) located in the subcutaneous tissue of the forearm or trunk. [sup][2] While they may be seen anywhere in the body, they are rarely found in the head and neck region. Intraosseous angiolipomas are even more exceptional. We describe the second reported case of angiolipoma of the skull and the first of angiolipoma of the frontal bone in a 16-year-old female.

CASE REPORT

A 16-year-old female patient was referred to the Department of Neurosurgery at our hospital with the complaint of swelling on the right side of the frontal bone with headache for a long time. Her neurological examination revealed no findings. She had no systemic diseases or a history of traumatic episodes. A computed tomography (CT) scan revealed a well-defined hypodense-lytic lesion, measuring 2 cm in diameter in the frontal bone [Figure 1]. Density measurement showed -80 HU within the lesion. No bone destruction or soft tissue component was seen. Radiological differential diagnosis included lipoma, hemangioma, Langerhans cell histiocytosis, epidermoid cyst and metastasis. She then underwent a frontal craniectomy and cranioplasty. In the gross examination, we observed a disk-shaped bone measuring 2.5 cm in diameter and a cavity in the middle of the bone 2 cm in diameter. When we sectioned the bone, a yellow-brown solid lesion was seen at the periphery of the cavity. The outer and inner cortical surface was intact.

On microscopic examination, the lesion was seen to have a widely infiltrating appearance between bone trabeculae with cortical bone erosion [Figure 2]a] and two components: Mature adipose tissue and blood vessels of variable size. The lesion had a variable appearance comprised of a predominantly vascular structure or adipose tissue [Figure 2]b and c]. Blood vessels were mostly thin-walled and rarely thick walled and abnormal vessels were mainly located at the periphery of the lesion. Most lesional vessels had a dilated appearance containing erythrocytes. In some of them, there were neutrophils filling the lumen and fibrin trombi [Figure 3]a]. The specific appearance of fibrin thrombi were showed in Masson Trichrom stained slides [Figure 3]b]. There was no mitosis and necrosis. Numerous mast cells were seen in the lesion. We counted an average of 10 mast cells per 1 high power field (HPF) in toluidine blue stained slides (in randomly selected 10 HPF) [Figure 2]d]. Immunhistochemically, the endothelium of lesional vessels was stained with endothelial markers (CD34, CD31) [Figure 4]a]. Vascular endothelial growth factor (VEGF) was positive in mast cells and the endothelium [Figure 4]b]. Mast cells were also stained with transforming growth factor beta (TGF-[sz]) and tumor

necrosis factor alpha (TNF-a) [Figure 4]c and d]. Thus, on histopathological examination, the lesion was diagnosed as an angiolipoma. A follow-up after 1 year did not show any complication or recurrence.{Figure 1}{Figure 2}{Figure 3}{Figure 4}

DISCUSSION

The term angiolipoma was described in 1960 by Howard and Helwig. [sup][3] However, the first case of intraosseos angiolipoma was reported by Polte et al . in 1976 in the left body of the mandible and presented with hyperesthesia of the lower lip and chin. [sup][4] Since then, only seven cases of intraosseos angiolipomas have been described: Four in the mandibula, two in the rib, and one in the parietal bone of the skull. [sup][1],[4],[5],[6],[7],[8],[9] Here, we describe a second angiyolipoma case of the skull and the first angiolipoma of the frontal bone. The clinicopathological features of previous reported cases are summarized in [Table 1].{Table 1}

Angiolipomas are known to have a different morphological entity from lipomas. They are very rarely 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 prominent characteristics: Presence of 50% mature adipocytes, interspersed angiomatous proliferation in the tumor, fibrin thrombi and presence of numerous mast cells, absence of nuclear pleomorphism, mitoses or necrosis, absence of other mesenchymal elements (smooth muscle, neural tissue) in contradistinction to other lipomatous lesions such as angiomyolipoma, angiofibrolipoma, angiomyxolipoma, liposarcoma, and hemangiomas. [sup][10],[11] 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 little lipomatous tissue and no fibrin thrombi. Vascular lumens of conventional lipoma have not fibrin thrombi. In our case, we described lesions consisting of two predominant components, >50% mature adipocytes and interspersed vascular structures, which are thin and thick walled, have dilated appearance containing erythrocytes, neutrophils filling the lumen some of which have characteristic fibrin thrombi and numerous mast cells in the stroma. We also noted a widely infiltrating appearance between bone trabeculae and microscopic cortical bone erosion. Thus, on histopathological findings, the lesion was diagnosed as an angiolipoma.

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. [sup][3] Some reports suggest that trauma is a causal agent. [sup][9] However, no history of trauma is seen in most angiolipomas. Similarly, there was no history of trauma in our case, while it must be noted that the patient was an adolescent female. In addition, fatty degeneration in hemangiomas and vascular proliferation in congenital lipomas have been implicated as possible predisposing factors. [sup][11]

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 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 [sup]2) [sup][12] We counted an average of 10 mast cells per 0.22 mm [sup]2 (45 mast cells per mm [sup]2) in our Toluidine Blue stained slides. Mast cells strongly produce VEGF, which is known to be essential for endothelial cells in angiogenesis, and TNF-a, which promote inflammation. TGF-[sz], also a regulator of angiogenesis, is a potent chemotactic factor for tissue mast cells and is released by mast cells, with tryptase which stimulates the proliferation of human vascular endothelial cells in human tumors. [sup][13] In this report, we demonstrated that VEGF and TNF-a were produced by plentiful infiltration of mast cells. Therefore, the result indicates that mast cells and pro-inflammatory cytokines produced by mast cells, including VEGF, TGF-[sz], and TNF-a might be responsible for the molecular mechanism of angiolipoma.

The symptoms of intraosseos angiolipoma vary depending on the location and size of the tumor. The most common first symptoms are swelling, hypoesthesia, and headache in the reported cases. [sup][1],[4],[9] However, three of the reported cases in the literature were symptomless. [sup][5],[6],[7] Consequently, the tumor may remain asymptomatic for many years. Our patient applied to our hospital with swelling and headache. Following examination, a single lesion was found in the frontal bone of the skull that reminded of Langerhans cell histiocytosis, which has a lytic appearance on the CT scan. Furthermore, the radiological appearance of angiolipomas resembles lipoma, hemangioma, Langerhans cell histiocytosis, epidermoid cyst, and metastasis. [sup][1],[14] Usually, these lesions are hypodense on CT and surgical excision or radiological follow-up needs to exclude metastasis. Histopathological evaluation is mandatory for final diagnosis. In the present case, there is no bone destruction or soft tissue component. Density measurement of the lesion revealed -80 HU which is consistent with the presence of adipose tissue. Differential diagnosis can be narrowed on lipoma, hemangioma or angiolipoma on the basic of radiological findings. [sup][13]

Clinical manifestations of Langerhans cell histiocytosis vary from isolated bone lesions to multisystem disease. Solitary bone lesion may be treated only through excision or limited radiation. The bone lesion should be excised for the final diagnosis. In the case, that histopatological evaluation was confirmed Langerhans cell histiocytosis, both of radiological and clinical long follow-up are necessary. On the other hand, complete surgical excision is the main treatment of angiolipomas and believed to be curative; no further treatment is needed in these lesions. [sup][1],[2]

CONCLUSION

To the best of our knowledge, only one case of angiolipoma localized in the skull has been previously described. Our case is the first known angiolipoma of the frontal bone. The diagnosis of angiolipomas of the bone must be kept in mind in radiological preoperative differential diagnosis of these types of lytic bone lesions.

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