Angiomyolipoma in the Temporal Region: A Case Report

Calenthia Ekawati,¹ Nico Lumintang,² Sherly Tandililing²

¹Specialist Study Program of Department of Surgery, Faculty of Medicine, Universitas Sam Ratulangi – Prof. Dr. R. D. Kandou Hospital, Manado, Indonesia
²Department of Head and Neck Surgery, Faculty of Medicine, University of Sam Ratulangi, Manado, Indonesia
Email: calent87@gmail.com
Received: April 11, 2023; Accepted: October 9, 2023; Published online: October 12, 2023

Abstract: Angiomyolipomas are benign tumors (consisting of fat tissues, smooth muscles, and blood vessels) that are often found in the kidney or liver. However, they are rarely found in the head and neck region and require proper management. We reported a 13-year-old girl presented with a mass in the right temple since last year. Musculoskeletal ultrasound (US) and contrast-enhanced computed tomography (CT) scan showed a well-defined hypoechoic lesion in the right temporal subfascial region, with a suspicion of a hemangioma. The patient underwent resection of the lesion. The diagnosis of angiomyolipoma was confirmed through a histopathological examination. The patient was subsequently discharged on the third postoperative day and had no further treatment. In conclusion, extraperitoneal angiomyolipomas are rarely found, and the diagnosis is confirmed by histopathological examination. The management of temporal angiomyolipomas is tumor resection. Keywords: temporal angiomyolipomas; histopathological diagnosis; surgical treatment
INTRODUCTION

Angiomyolipomas (AMLs) are a group of tumors from the family of perivascular epithelioid cell tumors. These tumors originated from mesenchymal and consisted of perivascular epithelioid cells. Angiomyolipomas are the most common benign tumor found in the kidney and are associated with tuberous sclerosis complex (TSC); they consist of vascular endothelial cells, smooth muscle cells, and fat cells. Generally, angiomyolipomas are associated with a genetic syndrome also caused by the germline mutation inactivating TSC1 and TSC2 genes.1,2

Angiomyolipomas are more commonly found in women (2-4 times). In addition, the sporadic form of angiomyolipoma is most often found in older patients with a mean age of 60 years. Sporadic angiomyolipomas in patients under 20 years old only constitute 3.5% of all cases.2

Although most cases occur in the kidney, angiomyolipomas may be found in other locations in rare cases. Other locations reported in studies include the liver, lungs, intestines, oral and nasal cavities, and, occasionally, the skin. Renal angiomyolipomas are more frequent in women, while cutaneous angiomyolipomas are more frequent in men.3,4 Nearly all cases of angiomyolipomas are renal angiomyolipomas (99.7% of all AMLs), and only 0.3% constitute extrarenal angiomyolipoma.5 In this study, we reported a 13-year-old girl with angiomyolipoma in a rare location, which is the temporal region.

CASE REPORT

A 13-year-old girl presented with a mass in the right temple for the last year. The mass was painless and originally small in size but progressively grew. She denied any history of head trauma, fever, tinnitus, weight loss, and familial history. Musculoskeletal ultrasound (US) and contrast-enhanced computed tomography (CT) scan showed a well-defined hypoechoic lesion in the right temporal subfascial region, with a suspicion of a hemangioma. (Fig. 1 and Fig. 2) The patient underwent resection of the tumor (Fig. 3). The diagnosis of angiomyolipoma was confirmed based on a histopathological examination. The patient was subsequently discharged on the third postoperative day and had no further treatment (Fig. 4).

Figure 1. Musculoskeletal ultrasound

Figure 2. Head contrast-enhanced CT scan
DISCUSSION

Angiomyolipomas are benign tumors that are commonly found in the kidney. Although very rare, these tumors may also be found in other locations. Renal angiomyolipomas are often found in women, however, cutaneous angiomyolipomas are generally found in men (70%). The age range of patients is 2-77 years, with a peak incidence between 30-50 years (median 48 years). Location predilection other than the kidney includes the head (76%), extremities (22%), and abdomen (2%). Of head tumors, the most common location is the ear (62%), followed by the nose (19%), and forehead, chin, or eyelids (19%). Cutaneous angiomyolipomas have many similar histological features to renal angiomyolipomas found in patients with tuberous sclerosis. However, cutaneous angiomyolipomas are not associated with tuberous sclerosis, and generally, the typical skin lesion found in patients with tuberous sclerosis, such as hypopigmented macules and angiofibroma, is not found in patients with cutaneous angiomyolipomas.4,5,6 In our case, the patient had an angiomyolipoma in the temporal region.

Most patients are asymptomatic and only experience visible or palpable nodular lesions, which are generally slow-growing. In our case, the patient had a mass in the temple region for the last year; the mass was initially small but grew in size. In addition, our patient had no other symptoms. In line with this, the literature showed that the time range of growth was two months until 40 years (median 5 years). Several patients may experience fluctuations in tumor size over time or fluctuations associated with temperature changes. These are clinical manifestations of the tumor’s vascular component. Other symptoms may include pain caused by increased sensitivity due to the location or trauma and obstructive symptoms associated with specific areas, such as the nasal cavity.4

Angiomyolipomas are classified as sporadic and TSC-associated types. Sporadic AMLs are usually asymptomatic and slow-growing. In this condition, patients with small (<4 cm) sporadic AMLs usually undergo active surveillance. On the other hand, TSC-associated AMLs develop at a much younger age and tend to grow rapidly over time compared to the sporadic type.5

One of the studies regarding angiomyolipomas in the head region is a study by Kim et al7 in 2017. The study reported a 60-year-old man with a cancerous mass in the glabellar region, and the mass progressively grew over the last three years. In that study, there were no other abnormalities, such as systemic signs of tuberous sclerosis complex (hypopigmented macules, facial angiofibroma). The patient was treated with surgical excision and had no adverse effects or recurrence. In line with the study by Kim et al,7 the patient in our case report also underwent surgical excision and had no adverse effects or poor outcome. However, the age predilection in our patient is not in line with the age predilection of extrarenal AMLs, which is mostly occurred in older patients.

The diagnosis of AMLs is generally sufficient with a computed tomography (CT) scan. The presence of a region with an attenuation of under -10 HU is a reliable sign and might identify fat tissues. Generally, AMLs detected from abdominal CT scans can be seen as a well-delimited and localized tumor in the renal parenchyma with -30 HU. Based on the fat content, angiomyolipomas are classified into three subtypes according to the HU in CT scans: fat-rich AML (≤ -10 HU); fat-poor AML (> -10 HU); and AML without fat content (fat-invisible) (> –10 HU). The low-fat form may cause a diagnostic challenge since fat tissues can be distinguished between angiomyolipomas.
and renal cell carcinomas. In fat-rich AML, the classical features in ultrasounds are hyperechoic lesions with a posterior acoustic shadow. This hyperechoic feature is the result of tumor content, consisting of fats, blood vessels, and muscles. However, this echogenicity of the mass may not always be found. As the fat content decreases, the echogenicity of the lesion may also be reduced. This result is not in line with the ultrasound finding in our patient, in which a well-defined hypoechoic lesion was found in the right temporal subfascial region.

Based on the histological examination, angiomyolipomas are defined as a benign pleomorphic mesenchymal tumor consisting of three histological components, including mature adipocytes, interlacing fascicles or smooth muscle cells, and abnormal tortuous convoluted thick-walled blood vessels without elastic layers. These three components are required in the histopathological examination to establish the diagnosis of angiomyolipoma.

Despite having similar pathological findings, renal and extrarenal angiomyolipomas might be considered as different disease entities due to the difference in histopathological behaviour, clinical behaviour, and demographics. Renal AMLs are small to medium in size and have blood vessels thickened by epithelioid components, while extrarenal angiomyolipomas have large blood vessels and often without epithelioid components. Furthermore, renal AMLs are associated with TSC and may present with signs of TSC in 30 patients, whereas extrarenal AMLs generally do not present with signs of TSC (except for liver AMLs) and have an excellent prognosis.

There are various treatments for renal angiomyolipomas, including active surveillance, embolization, mTOR inhibitors, and ablation. However, literature regarding treatment in temporal angiomyolipoma is scarce. In our patient, we conducted a resection of the lesion, and the diagnosis of angiomyolipoma was established based on the histopathological examination.

CONCLUSION

We reported a rare case of a 13-year-old girl with angiomyolipoma in the temporal region who underwent tumor resection. Extraperitoneal angiomyolipomas are rarely found, and the diagnosis is confirmed based on the histopathological result.

Conflict of Interest

The authors affirm no conflict of interest in this study.

REFERENCES

4. Sanchez NG, Romay AAA, Luna EM, Rodriguez ALP. Cutaneous angiomyolipoma—a distinct entity that should be separated from classic angiomyolipoma: complete review of existing cases and defining fundamental features. JMIR Dermatol. 2022;5(3):e40168. Doi: 10.2196/40168