

Soft Tissue Angiomatosis: A Rare Cases Report

I Wayan JuliSumadi¹, J. S. Leni Manna², I.G. A Sri Mahendra Dewi³, Ni WayanWinarti⁴, Eka W⁵

Department of Pathology Anatomy, UdayanaUniversity School of Medicine, Sanglah General Hospital, Denpasar

Abstract: Angiomatosis is a benign diffuse proliferation of blood vessels, concerning a wide body segment in a continuous pattern, involving multiple layers of tissue extending vertically or horizontally. These diffuse lesions make it difficult for clinicians to determine the limits of tumor-free tissue during resection, causing a high rate of angiomatous recurrence. We reported case of angiomatosis in 10-year-old female patient. Patient complaint was a lump on the left thigh. On Physical examination found mass of the left femur region. Macroscopically showed a fairly large tumor mass, lesion with ill-defined, brownish patches and chewy consistency. On histopathology examination shown feature of randomly arranged blood vessel proliferation between the connective tissue of the dermis profunda, subcutaneous fat, skeletal muscle tissue, and fatty tissue around the muscle. Blood vessels consist of arteries, veins and capillaries, some blood vessels appear to form clusters composed of thin-walled and thick-walled arteries and veins and capillaries. Based on clinical data, imaging and histopathologic examination, the patient diagnosed as angiomatosis.

Keywords: angiomatosis, soft tissue tumor, femur.

1. Background

Angiomatosis is a rare condition of soft tissue abnormalities with diffuse proliferative characteristics of benign blood vessels with well-differentiated architecture, concerning wide body segments in continuous patterns, involving various layers of tissue extending vertically or horizontally across the muscle compartment on a single layer of tissue.^{1,2,3} We report one case of angiomatosis in soft tissue in 10-year-old woman on clinicopathology perspective. This is an interesting cases, because the incidence is rare, high recurrence rate and a fairly difficult diagnosis because it has many differential diagnoses.

2. Case Presentation

Woman, 10-year-old, Balinese came to Sanglah Hospital Denpasar with complaints of lump in left thigh since 2 years ago. The lumps growth slowly, and the patient also complaint of pain that worsen which is exacerbated by the activity, and decreases while inactivity, but there is no disruption to daily activities. There is no weight loss and no appetite decreases. On physical examination, the tumor mass in the distal femur sinistra region, that 20x15 cm in size and on the sinistra pedis 6x3 cm in size. Both with venektasi, did not appear discoloration, deformity or atrophy. the tumor were solid and warm on mass palpation.

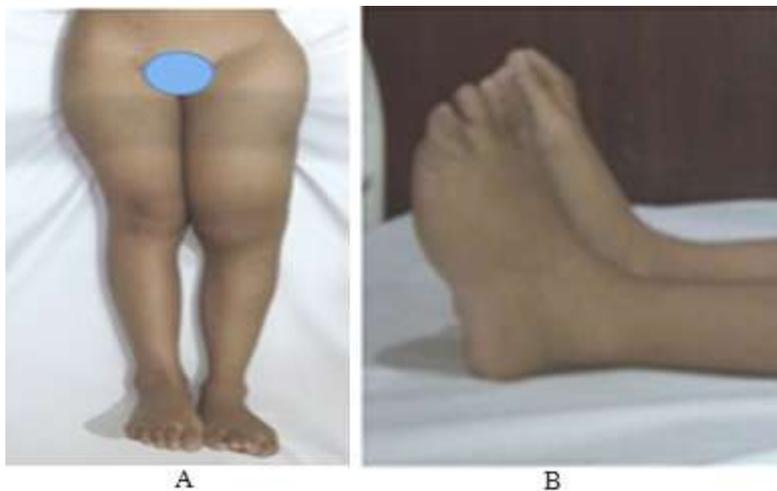


Figure 1: A) The tumor mass at distal femur sinistra, on the lateral region of femur B) Mass on the sinistra pedis.

X-ray photo in antero-posterior and lateral position of femur, genu and pedis sinistra shown that soft tissue tumor on genu region to 1/3 distal femur sinistra and lateral pedis sinistra. On MRI examination with and without contrast on femur sinistra shown an impression of tumor in the antero lateral femur sinistra, suspected angiolipoma. CT scan using of

contrast on femur sinistra, we obtained the impression of soft tissue bulging with heterogeneous lesions with fat components and hypervascularization therein which impressed reside in musculus vastus lateralis sinistra to subcutaneus.



Figure 2: Plain photo X-Ray on femur and pedis sinistra. A. Soft tissue tumor on 1/3 distal femur sinistra region. B. Soft tissue tumor on lateral pedis sinistra region.

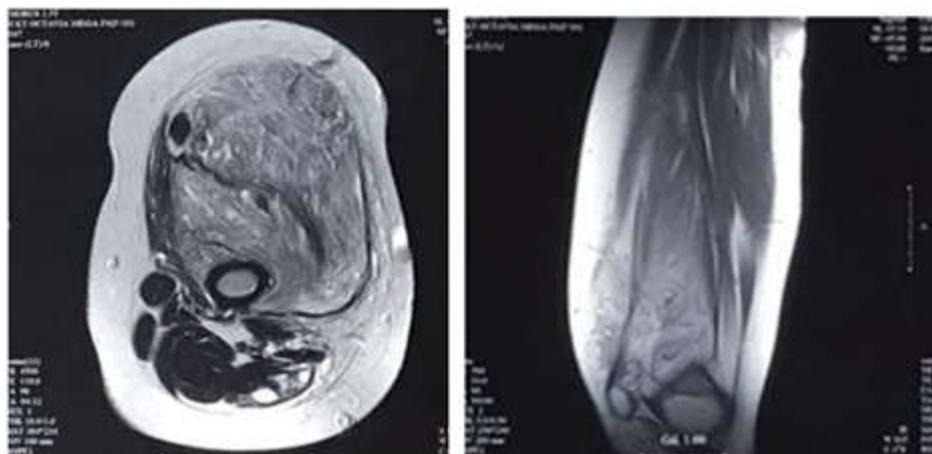


Figure 3: MRI result impressed there was tumor on anterolateral sinistra of femur, A) axial sinistra B) sagittal sinistra femur

Based on the anamnesis, clinical examination, CT Scan and MRI, the patient diagnosed with malignant soft tissue tumor of femur with differential diagnosis of benign soft tissue tumor. On January 18, 2017 core action and open biopsy performed by surgeon. We received pieces of tissue, the overall size of 2x1,2x0,4 cm with irregular shapes, brownish yellow and supple consistency on macroscopic examination.

Figure 4: Macroscopic finding result from core and open biopsy

On microscopic examination, pieces of tissue consisting of matured fat and fibrocollagen tissues. In some foci there were proliferation of blood vessels in various sizes between randomly arranged fat tissue. Some blood vessels are small, some larger with irregular walls, coated with an endothelial layer without atypia. In some focus also appears microthrombi. The conclusion is an angiomatosis, and the differential diagnosis is angiolipoma.



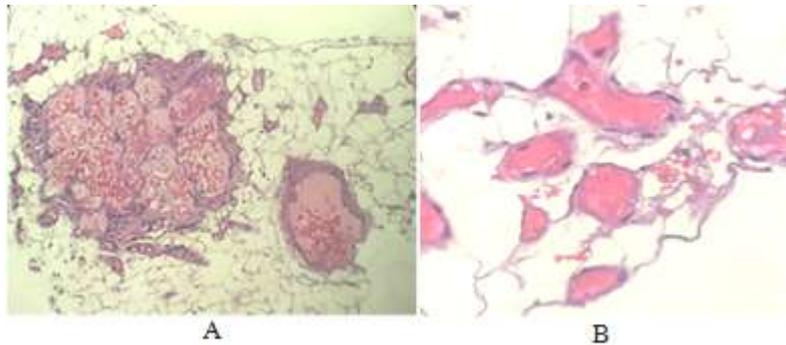


Figure 5: Microscopic finding. A. Blood vessels between mature fat (magnification 40x). B. Small vessels coated by endothelium (magnification 400x)

On February 10, 2017, the resection performed by surgeon. We received 2 resection tissues without lateralization with 18x12x4,5 cm and 7x3x1,5 cm in size. The smallest tissue appears covered by skin 5.5x1,5x0,2 cm in size. Irregular shapes, brownish yellow, supple consistency and by incision shown brownish patches appearance on macroscopic.

On microscopic examination, we found proliferation of blood vessels arranged randomly between connective tissue in the dermis profunda, subcutaneous fat, skeletal muscle tissue, and fat tissue around the muscle. Blood vessels consist of arteries, veins and capillaries, some blood vessels appear to form clusters composed of thin-walled and thick-walled arteries, veins and capillaries, partially veined with irregular walls. The entire blood vessel is coated with an endothelial layer without atypia. The limits of resection still indicate a vascular lesion. The conclusion is an angiomatosis.



Figure 6: Macroscopic tissue shown brownish patches after resection

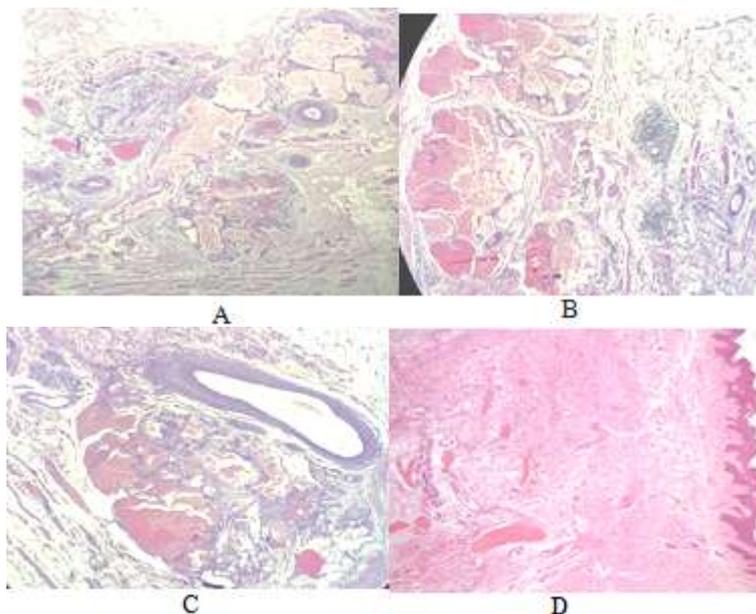


Figure 7: Microscopic finding. A to C: The proliferation of blood vessels arranged randomly, consisting of arteries, veins and capillaries. Some of which appear to form clusters composed of thin-walled and thick-walled arteries and veins (40x and 400x magnification). D. The skin tissue with proliferation of blood vessels in the dermis (40x magnification)

3. Discussion

According to the World Health Organization (WHO) definition, angiomas are a diffuse proliferation of benign blood vessels with well-differentiated architecture, concerning wide body segments in a continuous pattern, involving multiple layers of vertically extending tissue (skin, subcutis) and horizontally across the muscular compartment in one layer of tissue (about the various muscles).^{1,2} Histologically, the angiomas are a benign but clinically very extensive and is a serious blood vessel lesion due to high recurrence rates after initial excision. Angiomas are most common in the first two decades of life. Angiomas are slightly more common in females than males.^{1,2,3} Most lesions tend to be congenital, but due to their deep location, new lesions are clearly obvious in adolescence or early adulthood. Patients with angiomas present with persistent diffuse swelling and pain symptom.^{1,2} In our case, angiomas occurred in the first decades, 10-year-old woman with a lump in the left thigh and left leg and pain since 2 years. Angiomas considered as a malformation of the blood vessels, it appears in early life, growing proportionately consisting of inactive mitotic vessels, and showing no tendency for regression.^{1,4} More than half of cases occur in the lower limb, also at chest wall, stomach, and upper extremity.^{1,3} In some cases have been reported to occur in the mediastinum, head of the neck and vagina.^{5,6,7}

In macroscopic, boundary lesions are not firm, it is difficult to distinguish between tissues containing tumor mass and normal tissue. Size varies from small to large size.^{1,3} On the cross section, it look yellowish color with brownish spots. Based on the histopathologic examination, angiomas contains a mixture of large arteries, veins, and small capillaries. The most common pattern is the presence of thick and thin walled veins associated with a randomly scattered capillary between adjacent tissues. The veins have irregularly protruding walls and there is a branching cluster of venules and capillaries.^{1-3,5-9} The histopathologic features in this cases shown proliferation of randomly arranged blood vessels between the connective tissue of the deep dermis, subcutaneous fat, skeletal muscle tissue, and fat tissue around the muscles. Blood vessels consist of arteries, veins and capillaries, some blood vessels appear to form clusters composed of thin walled and thick walled arteries, veins and capillaries, partially veined with irregular walls. The entire blood vessel is coated with an endothelial layer without atypia. The limits of resection still indicate a lesion, so this case needs further clinical observation and evaluation to anticipate recurrence. Although biologically benign, nearly 90% of persistent lesions post-surgical excision, and 50% experienced multiple recurrences. The presence of tissues other than blood vessels involved in angiomas such as matured fat tissue, skeletal muscle and bone also cause some differential diagnoses. The differential diagnosis of angiomas includes arteriovenous malformation / hemangioma (AVMH), lipoma, angioliipoma, and liposarcoma.^{3,5-7} In this case the differential diagnosis can be excluded.

Arteriovenous malformation / hemangioma (AVMH) is a benign vascular lesion characterized by arterio-venous

shunts. Hemangioma has two variants, deep-seated and cutaneous form. When this hemangioma involves multiple tissues, the lesion is called angiomas. Hemangiomas are common in children and young adults. Its location is mainly in the head and neck with a clinical picture of pain and superficial cutaneous changes. On macroscopic examination, the tumor constraints are unclear, containing variations of large and small blood vessels, partially dilated. Histopathologic features are always correlated with clinicopathology and radiology. Hemangiomas are characterized by many blood vessels of different sizes, including veins and arteries. Areas resembling cavernous and capillary hemangiomas are common, including thrombosis and calcification. Fibrointimal thickening of the veins is a useful diagnostic clue. Local recurrence often occurs because of difficulties in complete excision of the tumor.

Lipoma is a benign tumor consisting of mature adipocytes. Conventional lipoma is the most common mesenchymal neoplasm and affects adults primarily in their 40s and 60s. Conventional lipomas may involve subcutaneous tissue or deep soft tissue or on bone surfaces. Intramuscular lipoma involves various locations including the trunk, head area, neck, upper and lower extremities and abdominal wall. Lipoma is usually present as a soft tissue mass without pain. Superficial lipoma is generally small (5 cm). Macroscopically lipoma, borderless, yellowish and shiny. In histopathological examination, conventional lipomas consist of mature adipocyte lobules. Lipomas sometimes have bone forming areas (osteolipomas), cartilage (chondrolipoma), or extensive fibrous tissue (myxolipomas). The cell ultrastructure has a single lipid cell with a core edge. The prognosis of lipoma is good and rarely relapsed. Angiolipomas are subcutaneous nodules composed of mature fat cells, mixed with small thin-walled capillary vessels, some containing fibrin thrombi. Angiolipomas are relatively common, usually appearing in late teens or early twenties. Angiolipoma is more common in men than in women. The most common sites are in the more common extremities, followed by the torso. Angiolipomas appear as small, multiple nodules, with soft consistency or pain. In macroscopic angiolipoma coated with hood, on the incision looks yellowish to reddish tissue. In a typical histopathologic examination angiolipoma consists of mature adipocytes and thin-walled capillaries that often contain thrombi fibrin. Vascularity is more prominent in the subcapsular region. Stromal spindle cells are more prominent in areas adjacent to blood vessels. The relative proportions of adipocytes and blood vessels vary. Angiolipomas are always benign and no tendency to recurrence.¹

Liposarcoma is a common sarcoma in adults and there are several subtypes that are histologically, biologically, cytogenetically, and molecularly analyzed, different from each other. The World Health Organization (WHO) divides into four subtypes: atypical lipomatous neoplasm / well-differentiated liposarcoma (ALN/ WDL), mixoid / round cell, dedifferentiated, and pleomorphic. A liposarcoma similar to angiomas is usually a liposarcoma mixoid. Mixoid liposarcoma occurs at a younger age with peak incidence in the fifth decade. These tumors often occur in

the lower extremities, especially the thigh area of the media and poplitea. Macroscopically this tumor is firmly bound, multinodular, and in pieces like jelly and shiny. In histopathologic examination, liposarcoma mixoid has a nodular growth pattern with increased cellularity at the edges of the lobules. There is a uniform mixture of non-spherical non-lipogenic cells up to oval and small signet lipoblast ring cells in prominent promotoid stroma, rich in fine capillary blood vessels with arborizing and chicken wire features. Management of angiomatosis is resection of tumor mass, but because of the difficulty in determining tumor mass limits, the recurrence rate of angiomatosis is very high.

4. Conclusion

Angiomatosis is a diffuse proliferation of benign blood vessels with well-differentiated architecture, which often occurs in adolescents or young adults. Clinical features of the mass or lump in the extremities, especially in the lower extremities. Morphological characteristics contain a mixture of large arteries, veins, and small capillaries. The most common pattern is the presence of thick and thin-walled veins associated with a randomly scattered capillary between adjacent tissues. cases have an appropriate picture for an angiomatosis. The recurrence rate of angiomatosis is very high because of the difficulty of determining resection limits on its management.

References

- [1] Weiss, S. W. 2013. Vascular tumour, angiomatosis. In : Fletcher, C. D. M., Bridge, J. A., Hogendoorn, P. C. W., Mertens, F., editor. WHO Classification of Tumours of Soft Tissue and Bone. 4th edition. Lyon: International Agency for Research on Cancer. p. 143.
- [2] Weiss, S. W. Goldblum, J. R. 2008. Benign tumors and tumor like lesion of blood vessels, Angiomatosis (Diffuse Hemangioma). In : Enzinger and Weiss's SOFT TISSUE TUMORS. Fifth Edition. Philadelphia: Elsevier. p. 665-666.
- [3] Lindberg, M. R., Lugas, D., Cassarino, D. S., Gardner, J. M., Stallings-Archer, K. 2016. Vascular Tumors (Including Lymphatic), Angiomatosis. In : Diagnostic pathology soft tissue tumors. Second edition. Philadelphia : amirsys, elsevier. p. 436-437.
- [4] Binesh, F., Aghili, K., Hakiminia, M., Vahidfar, M.R., Masumi, R. 2016. Disseminated Skeletal Angiomatosis Initially Misdiagnosed As Metastatic Tumor: A Case Report. Iranian Journal of Pathology, 11 (5): 443-447.
- [5] Pak, M.G., Choi, P.J., Choi, W.S., Kang, E.J., Roh, M.S. 2015. Mediastinal angiomatosis: a rare case report. J Thorac Dis, 7 (12): 662-666.
- [6] Shetty, S.R., Prabhu, S. 2009. Angiomatosis in the Head and Neck - 3 Case Reports. Head and Neck Pathol, 3: 54-58.
- [7] Grauso, F., Balbi, G., D'Aponte, M.L., Ronchi, A., Russo, R., Falcone, F., Messalli, E.M. 2015. Vaginal angiomatosis: differential diagnosis of a rare case. J Turk Ger Genecol Assoc, 16: 189-191.
- [8] Brooks, J. S. J. 2004. Disorder of soft tissue. In : Milis, S. E. Editor. Sternberg's diagnostic survival pathology. Fifth edition. Philadelphia : walters klewer. p. 166-168.
- [9] Rosai and Ackerman's. 2011. Soft tissues, Tumors and tumor like conditions of blood and lymph vessels. In: Rosai and Ackerman's. Surgical Pathology, Rosai, Volume I, Tenth Edition, New York, Elsevier Saunders: p. 2150-2151.