



Case Report

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Angiomatosis – A case report in comparison with vascular malformation

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Abstract

Angiomatosis is a diffuse vascular lesion involving multiple tissue planes. Its infiltrative nature makes surgical removal difficult and has high recurrence rate. Histopathologically, consists of proliferating blood vessels within or adjacent to major vessels. Here we present a case report of angiomatosis with a clinical diagnosis as vascular malformation.

Keywords: Angiomatosis, Vascular malformation, Histopathological diagnosis.

INTRODUCTION

Angiomatosis is a clinically extensive benign vascular lesion of the soft tissue that is symptomatic during childhood^[1]. Lattes used the term angiomatosis for benign vascular malformations of infancy^[2]. Hajdu^[3] and Allen^[4] defined it as multiple, diffuse hemangiomas in infants.

Vascular malformations are present at birth and persist throughout life^[5] results from abnormal signalling during embryogenesis^[6]. Vascular malformations are structural abnormalities of the arterial, venous, capillary and lymphatic system^[7].

This article describes the case of a 56 years old female diagnosed with angiomatosis along with a description on comparison between angiomatosis and vascular malformation.

CASE REPORT

A 56 years old female patient presented with the complaint of swelling in the left lower cheek region since childhood with a history of a gradual increase in size of the swelling since childhood with no history of pain.

Extraoral examination, showed facial asymmetry at left lower third of the face extending from ala-tragal plane to the lower border of the mandible. Skin overlying the swelling was normal in colour and texture.

Intraoral examination showed bluish discolouration of the left buccal vestibule. On palpation, the swelling was non-tender, soft, fluctuant, afebrile, compressible measuring 3x3 cm in size with no evidence of any kind of discharge. Diascopy test showed blanching on pressure. A provisional diagnosis of vascular malformation was given.

Excisional biopsy was done under general anesthesia by elevation of mucoperiosteal flap. The lesion revealed a thin cortical bone which could be easily pierced by blunt instruments. On removal of thin bone lesion appeared like a blood filled hollow in the bone. The lesional tissues were removed as minute scrapings. There was severe bleeding which was arrested using electrocautery and debridement was done with Cornoy's solution.

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Gross specimen consisted of multiple soft tissue specimens which were black in colour, firm in consistency with irregular margins.



Figure 1: Large diffuse swelling seen on the left lower face involving left cheek and lower lip.

The specimen was fixed and processed. H & E section of the received sample showed skeletal muscle tissue with intervening stroma. Numerous blood vessels of varying sizes were seen intermixed among the muscle fibres. Few vessels were attenuated in nature. Adipose tissue along with neural tissue was seen in adjacent areas. The histopathological diagnosis of 'Angiomatosis' was given.

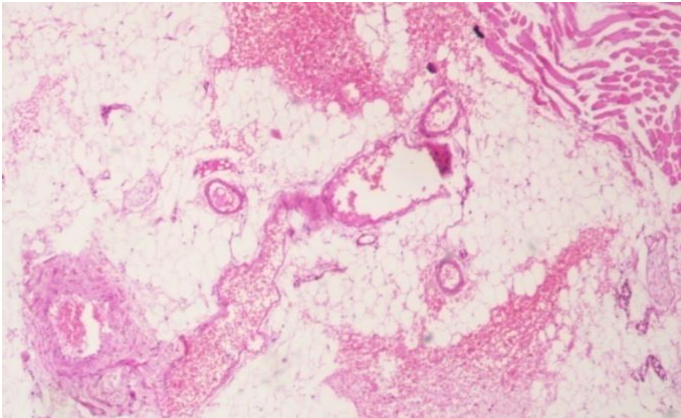


Figure 2: Photomicrographs showing clusters of capillaries with admixture of mature adipose tissue and muscles. (H&E, 10x)

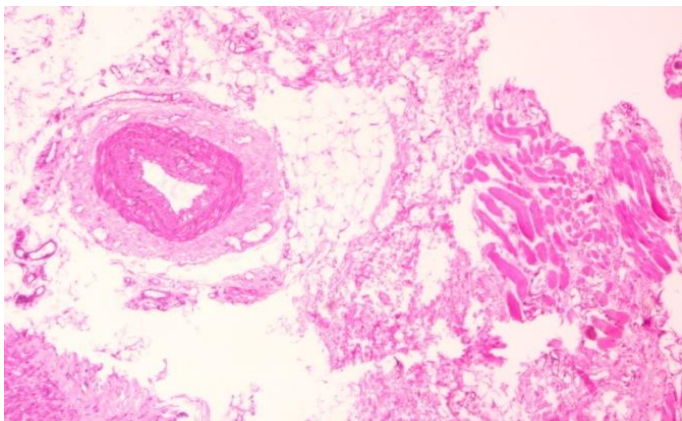


Figure 3: Photomicrographs showing admixture of thick walled blood vessel, mature adipose tissue and muscles.

On completion two small hole were made on the base of hollow bulb obturator. One opening was to pass the water to dissolve the salt and

second opening was to remove dissolved salt completely. These small hole were sealed with the self cure acrylic resin. The obturator was finished, polished and relined with soft liner and inserted in the patient mouth. (Fig.4) Post insertion instructions were given and patient was called for regular recall checkups which showed an improvement in speech, mastication, swallowing and facial esthetics.

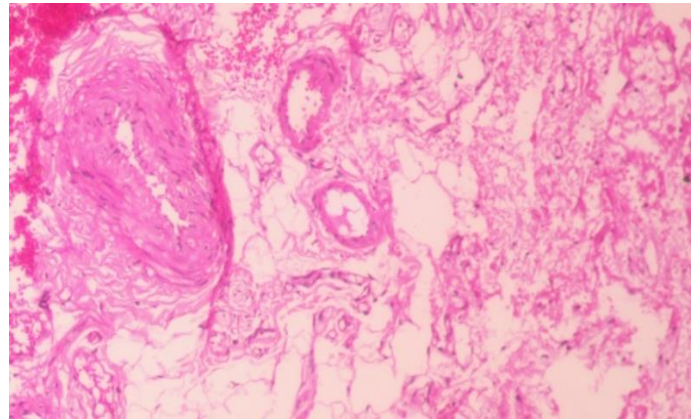


Figure 4: Photomicrographs showing thick and thin walled blood vessel.

DISCUSSION

Clinically the present case mimics vascular formation considering the age and clinical presentation. Histopathologically, vascular malformation shows progressively enlarging, dilated vascular channels lined by flattened endothelium, with vessels of uniform size. However, numerous blood vessels of varying sizes and thickness were seen intermixed among the connective tissue elements like fatty deposits and muscle fibres. The above features lead to the conclusion of angiomatosis rather than vascular malformation.

Angiomatosis can be described as a developmental anomaly characterized by diffuse blood vessel proliferation admixed with fat, muscle, fibrous tissue involving multiple tissue planes^[8]. Clinically it mimics hemangioma or arteriovenous malformation¹. It is also termed as infiltrative hemangioma^[9] diffuse hemangioma, haemangiomatosis^[10] and intra muscular hemangioma if muscles are involved predominantly. Rao and Weiss have suggested the term 'angiomatosis' that extensively involves different tissue types in a contiguous fashion in the body^[11]. Angiomatosis can be congenital or acquired. Acquired angiomatosis can be due to HIV or Bartonellosis. Congenital form may be sporadic or in association with such as Klippel Trenaunay syndrome, Gorham disease^[12]. However in our case no associated features of any syndrome were seen. Angiomatosis involves multiple tissue planes^[13] usually it occurs in the first two decades of life in childhood and adolescence with slight predilection for females usually with symptoms of diffuse persistent swelling sometimes associated with pain and discolouration. Very few cases are reported from other sites such as heart, abdominal wall, forearm, retroperitoneum and genitalia. Clinically, angiomatosis commonly presents as a bluish or reddish tint, firm, tender, ranging from 2 to 4 cm in size to the overlying dermis or mucous membrane^[14].

Histologically, angiomatosis shows large venous, cavernous and capillary sized vessels haphazardly scattered throughout soft tissue. The venous vessels are irregular and thick walls with occasional attenuations and herniations. In the second pattern, a central large vessel is surrounded by clusters of capillary sized vessels arranged in nodules diffusely infiltrate the surrounding soft tissue. The prominent amount of fat present in these lesions has led to use the term

infiltrating angiolipoma, suggesting angiomatosis to be a more generalized mesenchymal proliferation [1].

Vascular malformations are congenital anomalies present at any age [15]. Vascular malformations are thought to result from developmental errors by abnormal signaling processes during embryogenesis [16].

Vascular malformations are categorized based on their flow characteristics: fast-flow (arteriovenous malformation), slow-flow (venous malformation, capillary malformation and lymphatic malformation).

Capillary malformations (CMs) referred as “port-wine stain” appear at birth as macular pink stain and persist throughout life [17]. Capillary malformation are characterized by haphazardly arranged capillaries with flat endothelial cells, layer of pericytes and thin collagenous walls

Venous malformations appears clinically as bluish, soft, compressible tissue mass on head and neck, trunk and extremities. Venous malformation increase in size with puberty, hormonal changes, or infection [18]. Microscopically, composed of masses of veins of varying dimensions lined by a layer of endothelial cells.⁹ Phleboliths are pathognomonic of vascular formation with predisposition to thrombosis [19].

Histopathologically, venous malformation shows irregular venous type channels, with flat endothelial lining and lacks the smooth muscle of a normal vein. Luminal thrombi are frequently present, sometimes showing papillary endothelial hyperplasia.

Arteriovenous malformations (AVM) represent a arteriovenous shunting that develop through “nidus,” abnormal arterial and venous connection [20]. AVM is seen from birth but manifest in first or second decade of life. Clinically, AVMs appear bluish, not accompanied by pain and bleeding [21]. On palpations may be slightly compressible, soft and pulsatile swelling. It expands in response to trauma or puberty. Histologically, a mixture of thick walled arteries and veins, along with capillary vessels are evident. The syndromes associated are Capillary malformation-AVM syndrome, Bonnet-Dechaume-Blanc syndrome, Parkes-Weber syndrome [22]. Angiomatosis resembles many of the vascular tumors and malformations because of its infiltrative nature. Histopathological differential diagnosis includes infiltrating lipoma, angiolipoma, angiomylipoma and liposarcoma [23].

In the present case, complete resection is the preferred treatment of choice. The patient was followed up for a period of 6 months with no history of recurrence.

Hence the precise histopathological diagnosis is very important in treatment planning for the patient.

CONCLUSION

A possibility of angiomatosis was considered in comparison with the clinicopathological features of vascular malformation occurring in head & neck region. The precise histopathological identification of the uncommon entity and its differentiation from the more innocuous vascular lesion is important in the diagnosis.

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