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The Adipocerous Vascularity – Angiolipoma

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Preface Lipoma

Angiolipoma and hemangioma represent a spectrum of histological continuum, emerging as benign and gradually progressive neoplasia, wherein proportion of vascular and adipose tissue components can segregate the tumefaction. Normally, vascular fraction comprises of an estimated <10% of tumor mass within a lipoma, around 15% to 40% in angio-lipoma whereas hemangioma is almost comprehensively composed of aberrant vasculature with scanty intervening stroma [1]. Angio-lipoma is contemplated as a histological subtype of lipoma, constituting an estimated 5% to 17% of adipocytic neoplasia. Angiolipoma is a benign, encapsulated or non-encapsulated, adipose tissue tumor confined to the subcutaneous tissue, essentially comprised of mature adipocytes admixed with thin walled blood vessels and fibrin thrombi. In contrast to adjunctive lipomas, angiolipoma demonstrates extensive vascular proliferation and abundantly disseminated mature adipocytes [1,2]. Angiolipoma as a variant of lipoma was initially scripted by Bowen in 1912. Howard categorized angiolipoma as a contemporary neoplasm in 1960 on account of clinical and pathological distinction from lipoma [1,2].

Disease Pathogenesis

Hemangioma and lipoma configure a morphological continuum wherein an angiolipoma articulates an intermediate variant. Lipoma is a common mesenchymal neoplasm of adulthood and demonstrates an aggregation of mature adipocytes in the absence of cellular atypia [3]. Angiolipoma as an exceptional subcategory of lipoma displays mature adipocytes disseminated amidst a significantly vascular stroma. Hemangioma is an endothelial neoplasm preponderantly comprised of malformed vasculature. Extensively vascular, infiltrative and aggressive angiolipoma of the spinal cord represents a category of hemangioma. Despite histological concurrence and common mesenchymal genesis, aforesaid neoplasia demonstrates a markedly varied pathogenesis, especially within individual lesions or diverse subcategories. Notably, genetic aberrations discerned within cutaneous lipoma are absent in angiolipoma [3,4]. On account of various mechanisms of tumor engenderment, simultaneous emergence of divergent tumour types within a singular individual is pertinent, especially with familial instances. Concurrence of histological features indicate a natural progression of cogent neoplasia towards a singular subtype- either a gradual retrogression of vascular component of a hemangioma to configure an angiolipoma and eventually a lipoma or contrastingly, vascular metamorphoses of a lipoma with the articulation of an angiolipoma and thence a hemangioma [3,4]. Contingent to cytogenetic analysis, karyotype of angiolipoma is diverse from associated benign adipocytic neoplasia or lipoma, thereby indicating a hamartoma- like genesis. Factors implicated in the aetiology of angiolipoma are a precise history of a traumatic event, vascular metamorphoses of a lipoma and hormonal imbalance [3,4].

Disease Characteristics

Lipoma and angiolipoma are sporadic lesions although a familial incidence can be discerned. Familial angiolipomatosis appears as a component of familial lipomatosis and recapitulates concurrent neoplastic classification. Majority of hemangioma are sporadic although an autosomal dominant pattern of disease transmission is discernible. Nevertheless, morphological concurrence of lipoma, angiolipoma and hemangioma is infrequent [5]. Occurrence of multiple adipocytic neoplasia is indicative of angiolipomas whereas an isolated, singular lesion frequently enunciates a simple lipoma. Angiolipoma commonly appears in post pubertal individuals or young adults. The neoplasm is exceptional in children, middle-aged and elderly. An estimated 5% instances are familial [5]. Localization of angiolipoma within the spinal cord is extremely exceptional and accounts for approximately 0.04% to 1.2% of spinal neoplasia. Blood vessels within a spinal angiolipoma delineate an enlarged calibre, in contrast to cutaneous angiolipoma. Cutaneous angiolipoma is a distinct entity which commonly arises following second decade with possible implication of third generation family members [5,6]. Familial multiple angiolipomatosis is an infrequently delineated,

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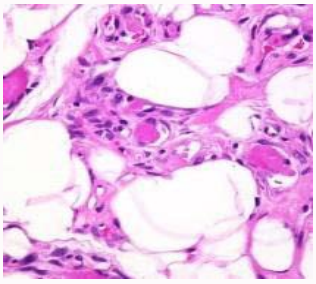

Figure 1: Angiolipoma depicting multiple aggregates of mature adipose tissue cells intermixed with thin walled vascular channels and extravasation of red cells [11].

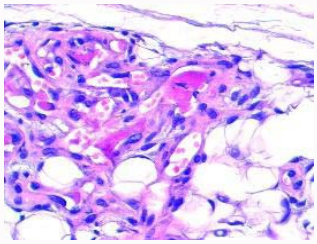

Figure 2: Angiolipoma demonstrating numerous blood vessels lined with thin endothelium, red cell extravasation and an admixture of mature adipocytes [12].

benign disorder demonstrating an autosomal recessive mode of disease transmission although certain instances depict an autosomal dominant mode of inheritance. Traditionally, familial multiple angiolipomatosis is concurrent with familial multiple lipomatosis [5,6].

Clinical Features

Angiolipoma is a gradually progressive, benign neoplasm composed of mature adipocytes admixed with aberrant, capillary sized vasculature. Angiolipoma is usually encountered as multiple, miniature, asymptomatic nodules below <2 cm magnitude [6]. Angiolipoma is further categorized into infiltrating and non-infiltrating subtypes wherein non infiltrating angiolipoma is a frequently occurring neoplasm engendered within younger individuals and manifests as a painless, subcutaneous nodule. Morphologic examination of non infiltrating angiolipoma depicts an encapsulated neoplasm comprised of mature adipocytes intermixed with thin- walled blood vessels. Infiltrating angio-lipoma arises in elderly subjects, is rarely encapsulated, infiltrates adjacent anatomic structures and commonly emerges in the head and neck region. Infiltrating angiolipoma depicts cogent clinical and histological criterion [4,6]. Clinical symptoms emerging from angiolipoma is pertinent to tumor localization. Subjects can demonstrate neurological symptoms such as weakness of lower limbs, gradually progressive numbness, dysesthesia on motility, a sense of imbalance, straining upon micturition and associated motor and sensory deficits. Familial instances display a cogent family history besides incrimination of several members [5]. Angiolipoma is frequently discerned as multiple, soft, painless or painful subcutaneous nodules situated upon the trunk, extremities or vertebral column. Angiolipoma is exceptional within maxillofacial region or gastrointestinal tract and can be determined with cogent morphology. Angiolipoma is usually cogitated within cutaneous tissue; an estimated 66% lesions appear upon forearm or chest wall although thighs or distal angiolipoma of spinal cord is commonly situated within the dorsal, midline, lower cervical region along ultimate lines of closure of rostral embryonic neural arch. Sudden onset of clinical symptoms such as weakness or sensory loss within the extremities can appear. Rapid progression of clinical symptoms is contingent to vascular engorgement, hemorrhage within the neoplasm, sudden thrombosis, altered calibre of incriminated capillaries or vascular steal syndrome [5,6]. On examination, tumour nodules are non fluctuant, non-tender, non-pruritic, well defined with an uninvolved superimposed epidermis [5].

Histological Elucidation

Macroscopically, reddish- yellow, friable, soft, extremely vascular neoplasm with intermingled spongy, hemorrhagic zones is delineated which melds with circumscribing, normal adipose tissue. Multiple enlarged blood vessels feed the neoplasm [7]. Alternatively, on gross examination, miniature, yellow-red, encapsulated, subcutaneous nodules beneath <2 cm dimension are discerned. Microscopy demonstrates fibrous and adipose tissue intermingled with numerous dilated blood vessels with a singular layer of flattened epithelium along with minimal foci of hyalinization [7]. On histology, a circumscribed, partially encapsulated tumefaction composed of mature adipocytes, capillaries and miniature blood vessels, a few of which are impacted with fibrinous microthrombi, is discerned. Tumor perimeter with aggregates of capillaries and mature adipocytes delineates entrapped normal parenchyma of incriminated organ. Aforesaid manifestation is associated with aggressive biological behavior with possibly enhanced tumour reoccurrence [7,8]. Angiolipoma exhibits aggregates of mature adipose tissue intermingled with branching capillaries and peripherally aggregated, thick walled blood vessels with demonstrable pericytes. Presence of hyaline or fibrin thrombi accumulated within the vasculature is a significant feature. Tumors with extensive cellularity are usually encapsulated; depict fibrin thrombi and distinct fibrous tissue septa. Mast cells can be discerned. Ancient lesions are imbued with fibrosis tissue. On ultrastructural examination, tumor cells display a decimation of Weibel-Palade bodies within the endothelial cells. Cytogenetic analysis almost always enunciates a normal karyotype [7,8].

Differential Diagnosis

Angiolipoma requires segregation from neoplasia such as Kaposi's sarcoma and angiosarcoma. However, aforesaid malignancies are devoid of adequate circumscription and tumor nodules are usually not confined to subcutaneous tissue. Tumor cells display cellular and nuclear atypia whereas tumour cell aggregates are devoid of scattered adipocytes [8,9]. Angiolipoma requires a demarcation from adjunctive painful nodules such as angioleiomyoma, eccrine spiradenomas, glomus tumor or traumatic neuroma. The neoplasm

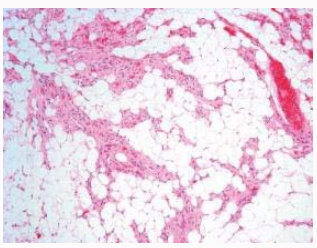

Figure 3: Angiolipoma demonstrating enlarged accumulates of mature adipose tissue cells with a prominent admixture of thin walled capillaries impacted with red cell extravasation and fibrin thrombi [13].

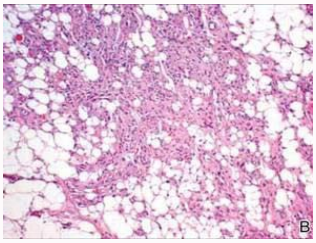

Figure 4: Angiolipoma delineating an extensive component of thin walled vasculature and commingled mature adipose tissue cells with erythrocytic extravasation [14].

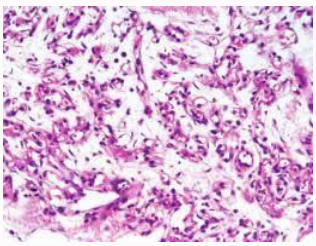

Figure 5: Angiolipoma depicting numerous capillary sized blood vessels impacted with red cells and an intermingling of adipose tissue cells [15].

can be a hemangioma comprising of aggregates of mature adipocytes, instead of neoplasia with an admixture of diverse mesenchymal elements. Infiltrative, intramuscular angiolipoma requires a distinction from intramuscular large vessel hemangioma depicting a replacement of skeletal muscle by adipose tissue [8,9]. Angiolipoma can be demarcated from associated lipoma or adipocytic neoplasia due to an excessive vascular element [8].

Investigative Assay

Magnetic Resonance Imaging (MRI) displays an enlarged, well encapsulated tumefaction which is hyper-intense upon T1 and T2 weighted imaging and is accompanied by multiple voids of the outflow tract. Fat suppression imaging displays decimation, thereby indicating an adipose tissue component of the neoplasm. Also, intense contrast enhancement is suggestive of an angiolipoma or arteriovenous malformation within cogent sites [8,9]. Hemangioma can concur within various locations. Simulating a simple lipoma, spinal angiolipoma appears hyper-intense on T1 weighted and T2 weighted imaging. Enlarged, hypo-intense foci within spinal angiolipoma emerging with non-contrast T1 weighted imaging can be correlated on account of enhanced vascularity. Majority of tumors are highlighted with administration of gadolinium whereas T2 weighted imaging can be variable although commonly appears as hyper-intense. Thus, an angiolipoma upon MRI delineates an intense, contrast enhanced image, which is absent in lipoma [8,9]. Adoption of Digital Subtraction Angiography (DSA) can confirm the presence of angiolipoma with emergent, multiple, adjacent blood vessels supplying the neoplasm. DSA can appropriately enunciate the vascular supply and aids demarcation of spinal angiolipoma from arteriovenous malformation [9]. On ultrasonography, a solitary, elliptical, well defined, isoechoic or hyperechoic, markedly vascularized lesion is demonstrated, separated from adjacent soft tissue with a well defined, hypoechoic capsule. Doppler scan demonstrates a venous outflow within peripherally arranged vasculature in the absence of intra-lesional blood flow, features which are suggestive of angiolipoma [10].

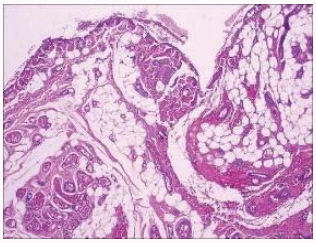

Figure 6: Angiolipoma displaying several thin walled, capillary sized blood vessels, fibrin thrombi, red cell extravasation and commingled mature adipocytes [16].

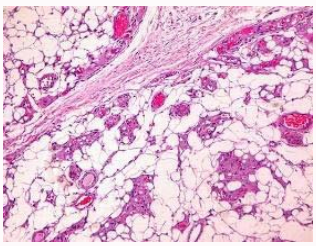

Figure 7: Angiolipoma depicting numerous capillary sized blood vessels impacted with erythrocytes, fibrin thrombi, fibrous tissue septa and admixed mature adipose tissue cells [17].

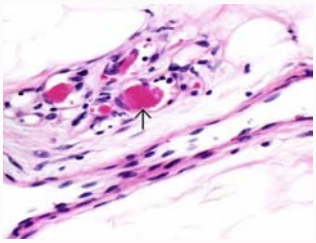

Figure 8: Angiolipoma exhibiting thin walled vasculature with red cell egress, attenuated endothelial lining and mature adipose tissue cells [18].

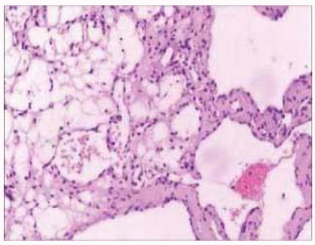

Figure 9: Angiolipoma enunciating branching capillaries and endothelium lined vasculature with intermixed mature adipose tissue cells [19].

Therapeutic Options

A comprehensive surgical extermination of the tumefaction is recommended. Preferential mode of therapy is total surgical excision of the nodule. Non-infiltrating angiolipoma depicts an absence of tumour reoccurrence following adequate surgical extermination, thus simple surgical eradication is curative [10]. Infiltrating angiolipoma is accompanied by an enhanced proportion of tumour reoccurrence, thus surgical extermination with a broad perimeter of normal parenchyma is recommended [10]. Postoperative Magnetic Resonance Imaging (MRI) can confirm satisfactory tumour eradication. Clinical symptoms are ameliorated immediately within the post-operative period. Vascular engorgement can concur as

Table: Histological features of non-infiltrating angiolipoma [3].

Emergence of 50% mature adipocytes constituting the neoplasm
Interspersed angiomatous proliferation in the tumour
Well encapsulated
Fibrinous micro-thrombi
Absence of other mesenchymal elements

intraoperative hemorrhage occurring during surgical eradication of the tumour [10]. Vascular embolization may not be indicated as vascular feeders are diffuse and normal vascular supply of the incriminated site may be jeopardized. Angiolipoma does not exhibit distant metastasis [10].

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