The Haemorrhagic Hinge—Synovial Haemangioma

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Abstract

Synovial haemangioma is an exceptional, benign, vascular neoplasm arising in association with joint cavities with pain and recurrent joint swelling. On histology, an admixture of distended vascular channels along with cavernous or capillary vascular articulations are denominated. Haemosiderin pigment deposits are accumulated along the synovial lining admixed with disseminated haemosiderin-laden macrophages. Synovial haemangioma mandates a segregation from conditions such as pigmented villonodular synovitis, synovial sarcoma, organizing haematoma, diverse arthropathies such as rheumatoid arthritis, juvenile chronic arthritis, haemophilic arthropathy, synovial osteochondromatosis, lipoma arborescens and cystic synovial hyperplasia. Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) are advantageous and recommended modalities for preoperative evaluation of the neoplasm. Cogent therapeutic options are vascular embolization, radiotherapy, laser induced arthroscopic abrasion and surgical extermination of the neoplasm as open or an arthroscopic procedure.

Keywords: Benign, Vascular, Joint.

Preface

Haemangioma of the bone comprises an estimated 1% of primary bone tumors. Soft tissue haemangiomas are infrequent neoplasms that arise within cutaneous or subcutaneous tissue wherein skeletal muscle and synovial tissue are exceptional sites of tumor emergence. Extensive, long-standing haemangiomas may be associated with concomitant lesions at diverse sites.

Synovial haemangioma is an exceptional, benign, vascular neoplasm arising in association with joint cavities, demonstrating pertinent clinical symptoms such as pain and recurrent joint swelling. Additionally, a benign, vascular lesion arising from synovium layered tissue as an intraarticular region, bursal space, or tendon sheath is denominated as a synovial haemangioma. The lesion may be contemplated as a variant of soft tissue haemangioma. Initially described by Bouchut in 1856, synovial haemangioma is predominantly intraarticular and is accompanied by delayed tumor discernment [1].

The vascular neoplasm usually occurs as an intermediate variety of hemangio-hamartoma or an arteriovenous malformation incriminating the synovial tissue with consequent intraarticular hemorrhage. Intraarticular and extraarticular haemangioma of the knee are exceptional neoplasms engendered from sub-synovial tissue. The condition may remain undetected and consequently untreated for significant periods. Antecedent therapy is preferred as synovial haemangioma can engender arthropathy on account of the
repetitive configuration of an intraarticular hematoma and infiltration of circumscribing skeletal muscle, adipose tissue, or cortical bone.

**Disease Characteristics**

Commonly, the knee joint is implicated besides the elbow and finger. The neoplasm is delineated in children and young adults. A male predominance is observed [2, 3].

Painful intra-articular haemangioma of the knee with hemarthrosis is classified into dual, exceptional categories which are challenging to treat and are denominated as

- **Synovial haemangioma and**
- **Arteriovenous malformation or hemangio-hamartoma**

Aforesaid lesions arise from the synovial membrane and can engender nontraumatic hemarthrosis. Arthropathy is initiated by synovial hemorrhage, intra-articular hemorrhage, or reoccurring mechanical activation [3, 4].

Bennet and Cobey categorized synovial haemangioma as

- **Localized, circumscribed, or pedunculated synovial haemangioma with sparing of the articular cartilage, minimal localized reoccurrence, and a favorable prognosis.**
- **Diffuse synovial haemangioma is progressive, challenging to excise with arthroscopic procedures, and demonstrates an enhanced possibility of reoccurrence [2].**

Synovial haemangioma is also subdivided by the site of tumor occurrence as

- **A juxta-articular variant which frequently depicts joint swelling**
- **An intraarticular variant which is typically associated with intraarticular hemorrhage**
- **Intermediate variant appearing as an amalgamation of distinctive variants [3, 4].**

The average age of disease onset in males is 12.5 years and in females is 10.9 years. The occurrence of intraarticular synovial haemangioma of the knee is exceptional in the pre-adolescent phase [3, 4].

Haemangioma emerging from tendon sheaths such as haemangioma of the tendon sheath may not be layered with true synovium or confined by synovial architecture and may not be designated as synovial haemangioma. Similarly, true synovial haemangioma or lesions arising in diverse regions such as the intramedullary compartment of bone, skeletal muscle, or subcutaneous tissue are excluded from the classification. Synovial haemangioma may represent a delayed stage of a post-traumatic lesion or a true, neoplastic vascular proliferation [3, 4].

**Clinical Elucidation**

Commonly a neoplasm of insidious onset, painful knee swelling with lack of history of trauma is observed in early childhood. Thus, a nontraumatic joint swelling associated with repetitive, hemorrhagic joint effusions and restricted joint mobility is exemplified. A spongy, compressible, palpable tumefaction is superimposed upon a joint [4, 5].

Subjects with skeletal maturity, recurrent joint effusion, and pain within the knee joint may delineate an intraarticular synovial haemangioma. Also, an extensive history of arthralgia or nontraumatic hemarthrosis may suggest an intraarticular haemangioma. Tumefaction appears as a soft, non-tender, elastic, painless, palpable, compressible, soft tissue mass of variable magnitude which enhances upon joint flexion [4, 5].
Tumefaction can be intraarticular as well as extraarticular. Synovial hemangioma of the knee depicts nonspecific features or manifests as hemarthrosis with localized pain, tenderness, swelling, and restricted joint mobility [4, 5].

Synovial hemangioma of the knee joint can extend from popliteal fossa to medial pouch epicondyle and articular surface with infiltration of the synovial layer of suprapatellar pouch. Of varying magnitude, tumefaction may extend from medial epicondyle to medial articular surface and adhere to the concave medial femoral condyle [4, 5].

Synovial haemangioma of intermediate type may originate from intraarticular synovium and extends from intraarticular region to extraarticular region [4, 5].

Nonspecific clinical representation may significantly delay disease discernment and cogent therapy. History of reoccurring, atraumatic, painless, hemorrhagic joint effusions is obtained. Relapsing hemarthrosis may ensue [4, 5].

**Histological Elucidation**

The intraoperative frozen section of excised synovial tissue depicts a haemangioma composed of mammoth, cavernous spaces admixed with capillary-sized vascular articulations. The lesion is designated as cavernous synovial haemangioma. Grossly, a discrete, intensely pigmented, brownish, soft tissue mass of varying magnitude with exuberant papillary fronds is observed arising from intraarticular synovium or adjacent bursa [5, 6].

On microscopy, an admixture of distended vascular channels along with cavernous or capillary vascular articulations is denominated. Haemosiderin pigment deposits are accumulated along the synovial lining admixed with disseminated haemosiderin-laden macrophages. Lymphoid follicles, stromal giant cells, or morphologic features indicative of aggressive biological behavior are absent [5, 6].

Generally, a diffuse, intermediate haemangioma engendered from the synovial membrane or sub-synovial connective tissue is exemplified along with concomitant, irregular arteriovenous connections [5].

Morphologically, synovial haemangioma is subdivided as

- venous subtype
- lobular capillary subtype- frequently discerned
- cavernous subtype- frequently discerned
- mixed subtype composed of an admixture of cavernous and capillary subtypes
- arteriovenous subtype and
- sclerosing haemangioma subtype [5, 6].

An intermediate subtype of synovial haemangioma is engendered with a combination of diverse subcategories such as cavernous and arteriovenous haemangioma [5, 6].

Dysplasia of abutting femoral condyle may arise and is designated as a secondary modification engendered by tumefaction of extended duration. Nevertheless, periosteal reaction and cortical destruction are documented in around <5% instances [5, 6].
Figure 1: Synovial haemangioma depicting enlarged vascular channels lined with endothelium and imbued with red cells with an encompassing fibrotic, mildly inflamed stroma [9].

Figure 5: Synovial haemangioma exemplifying distended vascular spaces layered with endothelium and impacted with red cells surrounded by a loosely configured, fibrous tissue stroma [13].

Figure 2: Synovial haemangioma delineating vascular arrangements distended with mature erythrocytes, a coating of endothelial cells, and surrounding fibrotic stroma [10].

Figure 6: Synovial haemangioma enunciating compact vascular articulations with circumscribing, extensive fibrous tissue stroma [14].

Figure 3: Synovial haemangioma demonstrating vascular configurations dilated with red cell extravasation, layered endothelial cells, and an encompassing fibrotic stroma [11].

Figure 7: Synovial haemangioma delineating dilated, endothelium layered vascular channels with incorporated red blood cells and an enveloping fibrotic stroma [15].

Figure 4: Synovial haemangioma exhibiting distended vascular arrangements enmeshed within a fibrotic stroma [12].

Figure 8: Synovial haemangioma depicting extensive vascular articulations layered with endothelial cells, imbued with mature red cells and an encompassing fibrotic stroma [16].
Differential Diagnosis

Synovial haemangioma mandates segregation from conditions such as pigmented villonodular synovitis, synovial sarcoma, organizing hematoma, diverse arthropathies such as rheumatoid arthritis, juvenile chronic arthritis, hemophilic arthropathy, synovial osteochondromatosis, or lipoma arborescens. The distinction can be obtained clinically or with magnetic resonance imaging (MRI)[3].

Demarcation on the adoption of imaging modalities is necessitated from

- Cystic synovial hyperplasia, a condition which is accompanied by significant synovial cell hyperplasia with multilayering, configured cystic structures with papillary articulations layered with synovial cells, palisaded histiocytes, focal fibrinoid necrosis, and hemosiderin pigment deposits. Cartilaginous fragments may be embedded within the cyst wall. The lesion commonly demonstrates singular peripheral enhancement of the images [3, 4].
- Synovial osteochondromatosis is a lesion of variable cellularity comprised of nodules of mature hyaline cartilage accumulated within synovial tissue and joint spaces. Chondrocytes depict variable atypia or binucleated cellular configurations. Aggregates of chondrocytes articulate tumor lobules and incriminate the synovium. Accompanying arthritis is usually absent [3, 4].
- Pigmented villonodular synovitis (PVNS) is a neoplasm composed of mononuclear stromal cells infiltrating the synovium, extensively vascularized villi layered with plump, hyperplastic synovial cells, an admixture of haemosiderin- laden macrophages, multinucleated giant cells, pigmented foam cells, or lipid-rich histiocytes, and frequent mitosis.
- Organizing hemorrhage is constituted by distended vascular configurations, fibrin deposition, focal hemorrhage, fibrosis, hyalinization, and neovascularization of subepithelial tissue [3, 4].
- Lipoma arborescens enunciates hypertrophic villous projections composed of mature adipose tissue layered with synovial cells and a variable intermingling of inflammatory cells [3, 4].
- Synovial sarcoma demonstrates a uniform, fascicular, biphasic tumor configuration composed of fibroblast-like, spindle-shaped cells admixed with gland-like epithelial structures and mucin containing glandular lumina. Distinctive epithelial component depicts cells with moderate, amphophilic cytoplasm and round to ovoid nuclei. A majority (90%) of neoplasms are immune reactive to cytokeratin (CK) or epithelial membrane antigen (EMA) and 60% to 70% of tumors react to CD99(MIC-2). The neoplasm is immune nonreactive to S100 protein [3, 4].

Apart from intraarticular neoplasms, segregation is required from diverse pathologies such as meniscal injury, discoid meniscus, meniscal cyst, disorders of medial patellar plicae, osteochondritis dissecans, juvenile idiopathic arthritis, and hemophilia [3, 4].

Investigative Assay

Intraarticular tumefaction generally demonstrates a non-infectious synovial proliferation that exhibits specific imaging characteristics.

The neoplasm is generally undetectable on plain radiographs which can be normal or nondiagnostic. Alternatively, plain radiographs can depict a soft tissue mass abutting the knee or incriminate joint, a soft tissue density suggestive of joint effusion, or a tumefaction that may be impacted with phleboliths or amorphous calcification, a pathognomonic feature. Plain radiographs may exhibit nonspecific modifications such as capsular thickening or bone erosion which is exceptional [7, 8].

An estimated below <5% tumefaction is associated with the periosteal reaction, cortical destruction, osteoporosis, advanced epiphysial maturation, a discrepancy in limb length, or arthropathy simulating hemophilia [7, 8].

Computerized tomography (CT) is a recommended pre-operative maneuver employed to ascertain bone
erosion or bone destruction. Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) are advantageous and recommended modalities for preoperative evaluation of the neoplasm. Magnetic resonance imaging (MRI) is a predominantly employed, accurate procedure associated with superior soft-tissue contrast which precisely defines the magnitude and extent of soft tissue tumefaction, associated chondral degeneration. The technique is preponderantly adopted for selecting optimal treatment strategies applicable for managing synovial lesions [7, 8].

MRI demonstrates a lobulated or diffuse, intraarticular mass or homogenous tumefaction of variable magnitude delineating minimal or intermediate signal intensity upon T1 weighted imaging. Significantly hyperintense background, possibly from blood accumulated within vascular spaces, lesion incorporated minimal signal intensity linear structures constituted by fibrous tissue septa or vascular channels are observed upon T2 weighted imaging. Contrast-enhanced T2 weighted imaging depicts significantly enhanced signal intensity. Fluid-filled channels may be observed [7, 8].

Medial femoral condyle with irregular margins and concavity may extend to the popliteal fossa. Intraosseous tissue is devoid of aberrant signals. MRI is generally indicative of a vascular, synovial neoplasm.

MRA displays a hyperintense region within the knee or incriminated joint, simulating the findings observed on MRI. Concomitant hyperintense region of varying magnitude abuts the lateral femoral condyle, indicative of extraarticular haemangioma. MRA is optimal for refining operative procedures and optimizing surgical measures to circumvent intraoperative hemorrhage. MRA can be beneficially adopted to assess diverse, comprehensive vascular neoplasms [7, 8].

The neoplasm can be adequately discerned by preoperative diagnostic arthroscopy and confirmed with cogent tissue specimens. Confirmation of nature, localization, and extent of tumefaction can be obtained with a precise arthroscopic examination. Additionally, modalities such as closed system venography or red blood cell scan can be utilized to determine the condition [7, 8].

**Therapeutic Options**

Synovial haemangioma is preferably treated in an antecedent manner as relapsing joint effusion with secondary chondral degeneration may ensue with delayed therapy. Prompt disease discernment and therapeutic intervention can circumvent a misinterpretation and progressive secondary degeneration within the joint. Delayed or absence of treatment of synovial hemangioma of the knee may engender dysplasia of medial femoral condyle [8].

The neoplasm can be suitably managed with diverse treatment methodologies. Cogent therapeutic options are vascular embolization, radiotherapy, laser-induced arthroscopic abrasion, and surgical extermination of the neoplasm as open or an arthroscopic procedure. Angiography can detect certain feeding vascular articulations which may be subjected to cogent vascular embolization. In the absence of embolization of pertinent vascular articulations, arthroscopic or arthrotomy-induced surgical eradication is the optimal treatment strategy [7, 8].

Comprehensive surgical excision of the tumefaction is an optimal therapeutic maneuver. Adequate surgical resection with a broad, unininvolved, tumor-free tissue perimeter is recommended in order to circumvent the reappearance of haemangioma. Alternatively, a subtotal synovectomy can be employed. Invasive, competent surgical resection is adopted to treat multifocal, extensive neoplasms [7, 8].

Singular arthroscopic management is optimal for miniature, localized neoplasms suitably assessed with cogent imaging procedures. Localized synovial haemangioma of extended duration and sparing of articular cartilage can be suitably managed with arthroscopic synovectomy [7, 8].
Vascular embolization may be imperative to manage a diffuse synovial haemangioma. Open surgical extermination of the neoplasm is optimal and recommended for treating enlarged lesions infiltrating into encompassing skeletal muscle [7, 8].

Synovectomy can be adopted for treating the circumscribing medial patella-femoral compartment. Encompassing soft tissue can demonstrate bluish discoloration and focal inflammation. Soft tissue may be cauterized to circumvent postoperative hemorrhage [7, 8].

Following adequate surgical excision, the asymptomatic tumefaction is devoid of reoccurrence or relapsing joint effusion [8].

Bone dysplasia is engendered on account of extended lack of therapy. Delayed commencement of suitable treatment produces osteoporosis, preliminary maturation of bony epiphyses, a discrepancy in limb length, and arthropathy simulating hemophilia. Aforesaid osseous manifestations may be circumvented with preliminary diagnosis and institution of therapy [7, 8].

References

10. Image 2 Courtesy: Libre Pathology
11. Image 3 Courtesy: Internet scientific publications
12. Image 4 Courtesy: Journal of hand surgery
13. Image 5 Courtesy: SS Journals
15. Image 7 Courtesy: Research gate
16. Image 8 Courtesy: Science direct

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